

LYMPHANGIOMAS OF THE OCULAR ADNEXA: AN ANALYSIS OF 62 CASES*

BY *Ira S. Jones*, M.D.**

LYMPHANGIOMAS around the eye are uncommon. Although they have been recognized and reported for nearly a hundred years, the reports, for the most part, have been based on a small number of cases or a limited observation. It seems worthwhile, therefore, to place on record an analysis, based on clinical and pathologic examination, of a substantial group of cases from two institutions, in order to delineate the features of lymphangiomas occurring in this region.

DEFINITION

Lymphangiomas are congenital, benign, slowly progressive tumors of the lymph-vascular system. The only one which is not benign is lymphangiosarcoma which has been reported in association with severe lymph stasis following radical mastectomy. The current naming of lymphangiomas is descriptive and includes capillary lymphangiomas, cavernous lymphangiomas, and cystic lymphangiomas. Some texts state that lymphangiomas are similar to hemangiomas and that a comparable type of lymphangioma exists for every hemangioma. Elephantiasis is a clinically descriptive name which undoubtedly includes many lymphangiomas but which likewise covers conditions without a disturbance of the lymph-vascular system. Some authors divide elephantiasis into congenital, which is presumably lymph-vascular, and acquired, which may be lymph-vascular on the basis of stasis of lymph vessels, or tissue edema related to inflammation and hyaluronic acid disturbance. Hemorrhagic lymphangiomas are those with a prominent element of hemorrhage into the lymph-vascular spaces. Hemangiolymphangiomas are those associated with hemangiomas.

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**From the Institute of Ophthalmology of the Presbyterian Hospital and the Department of Ophthalmology, Columbia University of Physicians and Surgeons, New York.

HISTORICAL REVIEW

CONJUNCTIVA

The first mention of a purely conjunctival lymphangioma was made by Morano in 1874 (59). Isolated cases were reported by Steudener (75), Imre (39), and Laskiewicz-Friedensfeld (47). A more detailed account with a presentation of three cases was made by Uhthoff in 1879 (80). Conjunctival lymphangiomas associated with hemorrhage were recognized and described by Leber in 1880 (48). Ottava's and Mules' observations (64, 60) of conjunctival lymphangiomas were followed, in 1890, by a report by Marcus Gunn (35) on a "lymphatic nevus" of the conjunctiva and other associated abnormalities in a ten-year-old girl with a lymphangioma of the conjunctiva. Gunn's patient also exhibited microphthalmos and a persistent pupillary membrane. Light-colored patches in the fundus on the affected side were interpreted as possible lymphangioma spaces within the globe. Deleccœuillerie (12), in 1892, wrote an exhaustive thesis on lymphatic cysts of the conjunctiva. In 1898, Sourdille (74) reported a case of congenital serous lymphangioma of the cystic variety involving the bulbar conjunctiva; Snell (73) described a solitary large clear cyst of the conjunctiva which appeared to be a lymphangioma; and Jocqs (41) published a dissertation in which he summarized the knowledge of lymphangiomas of the bulbar conjunctiva, and partially reviewed the literature up to that time. Zimmerman (88), in 1899, reported a single case of lymphangioma-hemorrhagica of the conjunctiva. An interesting gross anatomic description of a lymphangioma was furnished by MacCallan (52) who described thickening of the conjunctiva with tiny grapelike vesicles and pedunculated tumors. Further isolated case reports were made by Terson (78), Galezowski (28), Menacho (55), Knipe (44), and Libby (49).

In 1911, De Schweinitz (15), described an epibulbar growth which was red in color and appeared to be a hemangioma, but on microscopic examination no blood was present in the vascular spaces and it proved to be a cavernous lymphangioma. Erb (21) reported the case of a ten-year-old patient who had had a conjunctival lesion since birth. The gross picture indicated a hemangioma, but here, too, microscopic examination proved the tumor to be a lymphangioma. The use of X rays as a means of treatment was discussed by Franke in 1921 (26). In a paper reporting hemangiomas of the eyelid, Victor Ray also described a lymphangioma of the bulbar conjunctiva which had been present since the patient's birth and had grown slowly. When the patient was eight, the lesion was partially excised, but it recurred. It

was then treated with radium and it improved but was not completely eradicated.

In 1931, Schopfer (70) presented two cases of conjunctival lymphangioma, one of which was associated with lesions of the palate and buccal mucosa. Summarizing the knowledge of such lesions with special attention to the clinical appearance, he noted that the great majority of conjunctival lymphangiomas were on the bulbar conjunctiva and that faulty diagnosis often occurred because bleeding was common in the lymphangioma spaces. This led to the erroneous diagnosis of hemangioma. He pointed out that most of the lesions appeared during the early years of life and that the lesions must be differentiated from dermoids of the limbus. The possibility of congenital abnormalities, hyperplasia, and stasis in the origin or development of the lesions was discussed. He noted that complete surgical removal often was not possible and any portion which remained was subject to a gradual increase in size. The injection of alcohol and the use of irradiation were suggested as possible therapeutic agents.

Duke-Elder's *Textbook of Ophthalmology* (16) summarized what had been discovered about conjunctival lymphangiomas up to 1937. Assettati (3), in 1942, called attention to a combined capillary and cavernous conjunctival lymphangioma. A similar review of the subject of conjunctival lymphangiomas was published by Farnarier (23) in 1937, and Lopez Quinones (51) reported a case with a review of the more recent references in 1952. In a discussion of limbal tumors in 1948, Swann, Emmens, and Christensen (76) showed that limbal lymphangiomas could extend deeply with serious results. Kornblueth (45) reported a case in 1955.

LIDS

The first reference to lymphangiomas involving the lids was published, in 1863, by Von Graefe (32), whose patient had had a lesion in the left upper lid at birth. A description of lid lymphangiomas was given by Michel (58), writing in the Graefe-Saemisch *Handbuch* in 1876. Walzberg (82), in 1879, reported a lymphangioma, also present at the patient's birth, which involved the left upper lid and possibly the orbit and the side of the face. Logetschnikoff (50), in 1881, published an account of a rare case of elephantiasis of the lids, and the next year Teillais (77) published a report on a case of elephantiasis of the lids and conjunctiva present since birth. Similar cases were mentioned by Fage (22) and Gorand (30).

A review article summarizing knowledge of this subject was pub-

lished, in 1895, by Becker (6), who added one case of his own. A similar review appeared, in 1898, by Rombolotti (69), who used the term "elephantiasis" and stated that it might be congenital or acquired. He added one case to the literature. Additional cases were added by De Polignani (14), Van Duyse (19, 20), and Kroll (46). In 1902, Meyerhof (56) published a comprehensive review article of lymphangiomas of the conjunctiva and lids. He felt that solitary cysts might be almost stationary but that larger lesions could invade the orbit and the globe. The frequent association of similar facial lesions was pointed out. Excision was recommended as the treatment of choice although it was recognized that incomplete excision was followed by a recurrence. The electric puncture of the wall of a cyst was also suggested as a possible treatment. Uhthoff (80), in 1916, reported a case of tuberculosis of the conjunctiva of the upper lids combined with a lymphangioma. Von Hippel (37) reported three cases of cavernous lymphangiomas of the conjunctiva and lids in which diagnosis was hampered by the presence of bleeding into the lesion. Excision of the lesions in his cases was difficult because the bleeding obscured the limits of the lesion. Attempts were made to use carbon dioxide snow and alcohol injections to eradicate these lesions. Bonnet and Colrat (11) added a case of congenital lymphangioma of the lids to the literature in 1935. Heintz (36) added three more in 1943, and Grom (34) reported one which developed after the scratch of a cat. A capillary lymphangioma of the lid which ulcerated spontaneously was recorded by Bishay (8). Andersen and Asboe-Hansen (2) reported an elephantiasis involving all four lids which was treated with some improvement, by ACTH.

Duke-Elder's *Textbook of Ophthalmology* (17) summarized the knowledge of lid lymphangiomas up to 1952, and a comprehensive review of the subject of elephantiasis of the lids was made by Bobb (9) in 1955. Among the causes of elephantiasis, he listed neurofibroma, hemangioma, lymphangioma, leukemia, recurrent inflammation, filaria, and hyaluronic acid disturbance.

ORBIT

Although Wecker (83) reported a cavernous tumor of the orbit with recurrent exophthalmos in 1868, the credit for the first report of orbital lymphangioma is most often given to Von Forster (25). In a series of six primary tumors and three secondary tumors of the orbit, he presented one case of lymphangioma of the orbit in a 46-year-old man. Nettleship (61), in 1884, reported on a 15-year-old boy with a lymph-

angioma involving the conjunctiva, brow, and orbital cavity. Hemorrhage had occurred into the lesion from time to time, and a lamellar cataract was present in the eye on the same side.

In 1886, Wiesner (85) reported the case of a woman with one year of swelling of the right lower lid followed by double vision and a palpable tumor between the globe and the outer orbital rim. Microscopic diagnosis of the tumor after it was removed indicated that it was a cavernous lymphangioma. Wiesner discussed lymphangiomas of the orbit in general and concluded that they were congenital tumors although they appeared in an area where no lymph vessels had been demonstrated. The difficulty of differentiating lymphangiomas from hemangiomas in the orbit was mentioned and the intermittency of the exophthalmos was noted as a helpful clinical point. Wiesner suggested that orbital lymphangiomas might be a backward extension from lymph vessels of the conjunctiva.

In the United States Ayres (5) reported an orbital lymphangioma in 1895. In 1896, Silcock (71) presented the case of a 21-year-old woman with one year of proptosis of the left eye. At surgery an orbital cyst containing clear fluid was removed. Within five years the condition had recurred and again excision was required. After the second excision, heat coagulation was used, but again the cyst recurred. Microscopic examination of the tissue which had been removed led to a diagnosis of cavernous lymphangioma.

Wintersteiner (86), in 1898, reviewed the literature, mentioning the orbital lymphangiomas reported by Von Forster, Wiesner, and Ayres, and added a report on a 12-year-old child with an exophthalmos which had been slowly progressive since birth. He noted this exophthalmos was more pronounced when the eyes were inflamed and that pressure on the globe decreased but did not eradicate the proptosis. Exenteration of the orbit and examination of the tissue which was removed showed endothelial-lined spaces and a sparse connective tissue stroma, as well as typical lymph follicles in the stroma. Wintersteiner's analysis of the case indicated that the clinical diagnosis lay between hemangioma and lymphangioma. He felt that the presence of lymph follicles and of lymphatic fluid was diagnostically important, but that the presence of blood and blood pigment in many parts of the tumor was confusing and resulted from bleeding into the lymphangioma spaces at various times. He noted that hyaline degeneration of the endothelial cells and of the connective tissue wall was prominent and he felt the site of origin of the orbital lymphangioma was the perivascular lymph space of a posterior ciliary artery.

Knapp (43), in 1899, described five rare tumors of the orbit of which two appeared to be lymphangiomas. Two cases of cavernous lymphangioma affecting the face, lids, and orbit were reported by Herschberg (38) in 1906. He cited the progressive nature of the afflictions and the repeated surgical procedures necessary to cause improvement. A comprehensive review of lymphangiomas of the orbit was offered in 1908 by Knapp (43) who added the case of a 24-year-old woman with six years of progressive exophthalmos of the left eye. A mass was palpated in the orbit and there was blurring of the optic disc. During surgery the orbital mass was opened and found to contain clear fluid. The microscopic examination of a portion of the wall of the cyst showed cavernous spaces containing lymph, a fibrous stroma with blood vessels in the septae, and an endothelial lining to the cystic spaces. Knapp classified lymphangiomas as ectasia with hyperplasia, homoplastic neoplasia or endothelial proliferation, and heteroplastic neoplasia or formation of new lymph spaces. Electrolysis was suggested as a treatment for these lesions which should be subjected to trial.

In 1908, Fehr (24) added the case of a 45-year-old lady with exophthalmos to the literature. This patient had a history of progressive protrusion lasting a year and a half and at operation was found to have a tumor 27 mm. in its greatest diameter. Microscopic examination of the tissue showed it to be a cavernous lymphangioma. Dejonc (13) in the same year added two more cases and speculated as to the origin of orbital lymphangiomas. In 1912, Bogatsch (10) described a rare occurrence, a case of endothelioma lymphangiomatosum of the orbit. His patient was a 27-year-old man with exophthalmos on the left. This was said to have arisen following an inflammatory process. At operation a solid tumor was removed. The study of this tumor microscopically showed it to be an encapsulated mass with hyaline changes in the tumor and spaces of a cavernous nature almost solidly filled with proliferated endothelial cells.

In 1915, Mackay (53) reported the case of a 15-month-old child with exophthalmos and a bluish mass or cyst in the conjunctiva of the upper lid of the same side. There was no pulsation. The cyst was punctured and dark bloody fluid was removed. The proptosis was reduced but reappeared again in a few days. Surgical excision of a portion of the mass was followed by radiation treatment. After one year the proptosis again began to increase and X rays were again used. Four years later the proptosis recurred and X-ray treatment was again applied. The tissue which had been removed at surgery was found to

be a cavernous lymphangioma. The author stated that such lesions might disappear spontaneously in the course of a few months in childhood.

Gradle (31), in 1920, reported a cavernous lymphangioma of the orbit which had begun when the patient was six months old and which was excised when he was seven years of age. The lesion was a cyst containing old blood, but on microscopic examination it was found to be a cavernous lymphangioma. In 1922, Niosi (62) presented a case and a short discussion. Following a case report of an orbital lymphangioma in an eight-year-old girl, Franklin and Cordes (27) collected 13 cases of lymphangiomias of the orbit from the literature. Their own patient had had protrusion of a blind eye since the age of two. At the initial surgery an orbital cyst was found and drained of colorless fluid. Two and one-half months later the exophthalmos recurred and the cyst was carefully dissected out. Microscopic examination showed endothelial-lined spaces in a fibrous tissue stroma with some calcified bodies and thickened endothelium. The diagnosis was lymphangioma.

Nizetic (63), in 1925, presented a case of cavernous lymphangioma of the orbit characterized by progressive exophthalmos. In the same year Smith (72) reported on a ten-year-old girl with a gradual protrusion of the left eye and an orbital mass above the globe containing cystic fluid and blood. Following the initial surgery, the condition recurred until 12 mm. of exophthalmos were present. A second exploration revealed a cystic mass containing brown fluid located behind the globe. The muscle cone was involved and the mass extended laterally to the orbital walls. The orbit was partially exenterated. Examination of the specimen showed edematous connective tissue with lymph cell aggregations and cystic spaces containing organized blood clots. The connective tissue septa contained numerous blood vessels with thickened walls. The globe which had been removed was normal. There was thickened tissue around the optic nerve and it was felt that the lymphangioma extended along the nerve sheath of the optic nerve to the globe and ramified in the episcleral tissues. The patient received radium treatment in addition to the surgery.

Three cases of lymphangioma of the orbit were added to the literature by Meisner (54) in 1926. The author felt one case was congenital, the second inflammatory, and the third contained features of both. Birch-Hirschfeld (7) collected the available information about lymphangioma of the orbit in 1930. Wolff (87), in 1932, reporting a case of recurrent proptosis, suggested that the combination of proptosis with inflammation and with spontaneous subsidence and recrudescence

but without suppuration probably constituted a syndrome. His own patient had had these features over a period of years beginning at age four. At the onset of proptosis pain, vomiting, and elevation of temperature occurred. The proptosis lasted about one month at a time, during which time the inflammatory signs and symptoms gradually disappeared. When the patient was 11 years old, a palpable tumor appeared in the upper outer angle of the orbit, surgery was performed, and the tumor was found to be a cavernous lymphangioma. The author examined 20 cases of cystic lymphangioma of the orbit collected from the literature and felt that only a few resembled his case.

Michail (57) added a case to the literature. Jess (40), who added three more cases to the literature and analyzed the cases previously reported, stated that the natural history of lymphangiomas was similar to that of hemangiomas and that the lesions were present at birth and developed slowly but were prone after a long time of very slow progression to show sudden rapid changes. Wheeler (84), in 1937, reported orbital cysts without epithelial lining, both in two adult males with unilateral proptosis. Both had larger orbits by X-ray examination on the affected side and both at surgery revealed cysts of the orbit filled with old degenerated blood. Pathologic examination of both specimens revealed many similarities to lymphangioma of the orbit. Radnot (66) discussed the pathology of orbital lymphangiomas and added two cases to the literature.

Duke-Elder's *Textbook of Ophthalmology*, volume V (18), published in 1952, added up 30 cases of orbital lymphangioma beginning with Von Forster. Duke-Elder's view was that these lesions resembled hemangiomas and arose congenitally in association with the perivascular lymphatics of the orbit. He felt associated lesions on the lids, conjunctiva, and face were common, and that the proptosis was very slowly progressive throughout life. He noted 10 mm. of proptosis as the maximum recorded. Intermittency was also mentioned by Duke-Elder and was attributed to mild inflammatory attacks or more rarely to hemorrhages. Grignolo (33) added a case with facial involvement in 1953.

As cases of various lymphangiomas in the ocular adnexa appeared in the literature the descriptions were incorporated in standard textbooks of ophthalmology. The accounts indicated conjunctival lymphangiomas to be more common than those of the lid; those of the orbit were the rarest. The number of cases reported in the literature, however, is inversely proportional to the apparent incidence. Consequently,

it appears fruitless to make any estimate as to the number of conjunctival lymphangiomas which have been observed. With regard to lymphangiomas involving the lid, the situation is complicated by an obscure terminology. The use of the term elephantiasis of the lids has included lymphangiomas, but most of the cases appear to be placed on some other basis. A similar difficulty exists with regard to orbital lymphangiomas. Perhaps a few orbital blood cysts not related to lymphangiomas have been included in this diagnosis, but certainly many orbital blood cysts secondary to a lymphangioma cysticum have been ascribed to other causes. My own scrutiny of the literature has revealed 34 cases, prior to the present paper, which appear to be lymphangiomas of the orbit.

INCIDENCE

It has not been possible to determine the incidence of lymphangiomas from the literature. The present study is based upon 62 cases of which 34 were secured from among 9,000 successive ophthalmic specimens. This represents an incidence of 0.40 percent with respect to the accession list in this particular pathology laboratory. The number of lymphangiomas in general among all admissions to the medical center, of which this ophthalmic institution is a part, is approximately 140. If the population base for this institution is about one million people, and the length of time during which the cases have been collected is one generation, then the incidence of lymphangiomas is 14 per 100,000 population. Since the 34 lymphangiomas involving the ocular adnexa which are reported from this institution are a portion of the 140 total lymphangiomas, the incidence of lymphangiomas involving the ocular adnexa works out to 3.4 per 100,000 population. The regional distribution of lymphangiomas in this institution is as follows:

REGIONAL DISTRIBUTION OF LYMPHANGIOMAS

	<i>Percentage</i>
Scalp and face	20
Lids, conjunctiva, and orbit	19
Neck	15
Chest and back	13
Leg, thigh, and foot	10
Tongue and palate	8
Arm and hand	6
Abdomen	2
Anus and rectum	2
Other sites	3

The distribution of lymphangiomas involving the ocular adnexa is as follows:

STRUCTURES INVOLVED BY LYMPHANGIOMAS ABOUT THE EYE

<i>Structures</i>	<i>Approximate percentages</i>
Conjunctiva	33
Orbit	25
Lids	8
Lids and conjunctiva	6
Lids, conjunctiva, and orbit	5
Conjunctiva and orbit	4
Face and lids	2
Face, lids and conjunctiva	2
Face, lids, conjunctiva, and orbit	6
Face and orbit	2
Face, lids and orbit	4

PRESENTATION OF CASES

LYMPHANGIOMAS OF THE CONJUNCTIVA

CASE 1. Age, 22 years. For four months there had been dilated vessels in the conjunctiva of the right eye temporally. The patient had had a blood vessel cut to limit the blood supply of this area without effect. Examination showed a pale watery mass on the bulbar conjunctiva of the right eye, extending from the temporal fornix at the canthus to the insertion of the external rectus muscle. A rich blood supply was apparent. With the slit lamp, dilated tortuous capillary channels filled with a clear fluid could be seen. Pigment granules could also be seen with the slit lamp. The diagnosis was lymphangioma of the conjunctiva. Excision and plastic repair were carried out in the operating room. The microscopic description of the specimen was as follows: "The conjunctiva shows thickened irregular epithelium and in one area intra- and subepithelial nest of cells of a nevoid character. In the subepithelial area are collections of lymphocytes and many small endothelial-lined vessels. In one area there is a cyst lined with flat endothelial cells and containing cellular debris." This cyst was a lymphangioma associated with a nevus (Figure 1).

CASE 2. Age, 57 years. The patient gave a history of a bloodshot area on the right eye for eight years which had cleared from time to time. Examination showed a localized swelling of the bulbar conjunctiva between six and nine o'clock. This was elevated and watery to flesh-colored. A provisional diagnosis of lymphoma of the conjunctiva was made. A local excision of the suspicious lesion was carried out. Microscopic examination of the tissue which was removed revealed numerous small blood vessels and edematous

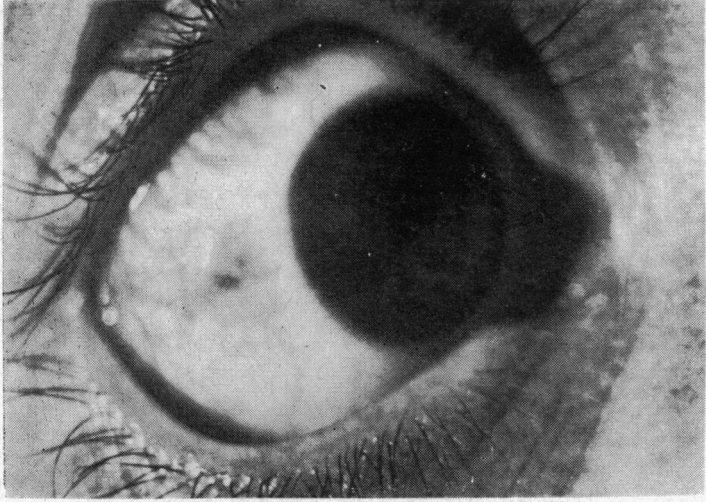


FIGURE 1. CASE 1

Lymphangioma of bulbar conjunctiva associated with a pigmented nevus.

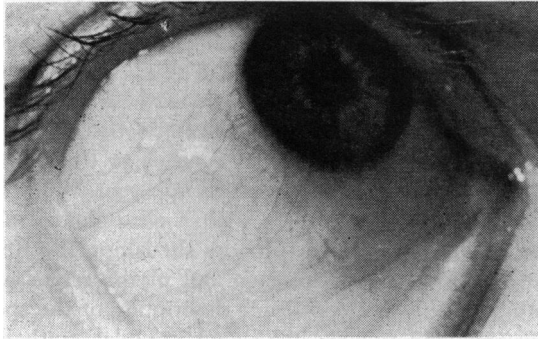


FIGURE 2. CASE 2

Lymphangioma of bulbar conjunctiva present eight years.

tissue containing many endothelial-lined convoluted spaces. The diagnosis was lymphangioma of conjunctiva (Figure 2).

CASE 3. Age, 29 years. At the time of admission the patient had had a tender red left eye for twelve years. This eye had undergone surgery three times for a cyst of the conjunctiva about ten years previously. The redness and tenderness would clear for a while and then return. The inflammation present at the time of admission had lasted for about two and one-half weeks. This blemish was hampering the patient's career as an actor. The earliest symptom which the patient had noted, 12 years previously, was a

protrusion of the inner aspect of the left eye. When it became painful, a cyst had been diagnosed and this had been treated first with an electric needle and then by attempted excision. However, within two years the condition had returned. Prior to the present inflammatory episode the eye had been clear for six to eight months. The examination showed an elevated lesion occupying the region of the caruncle and the semilunar fold in the left eye. This was highly vascular and angiomatous in appearance. It extended to the cul-de-sac above and below and showed adhesions to the lids both above and below. Some scar tissue was present. There was no exophthalmos. The clinical diagnosis was lymphangioma of the conjunctiva. An excision of the mass together with affected conjunctiva was carried out. A plastic repair was instituted. Microscopic examination of the tissue which had been removed showed a gross specimen $7 \times 4 \times 3$ mm. It consisted of epithelium and subcutaneous tissue, in the deeper layers of which there were numerous endothelial-lined spaces varying markedly in size. Many of the spaces were empty, but some contained blood. This was a cavernous lymphangioma with a small but definite lymphocytic element.

CASE 4. Age, 54 years. When this patient was first seen, he had had an irritation of the left eye for several days. A lymphangiectatic-appearing growth was present on the bulbar conjunctiva temporally. Excision was carried out under local anesthetic. Microscopic examination of the specimen showed many large cavernous spaces lined with endothelial cells and containing an eosinophilic homogeneous substance. The diagnosis was cavernous lymphangioma.

CASE 5. Age, adult. This patient had a cavernous lymphangioma of the semilunar fold. A cystic growth had been noted on the semilunar fold 18 months before admission. It had been partially excised twice but refilled and reconstituted itself almost immediately. A third and complete excision was carried out and microscopic examination of the tissue revealed a cavernous lymphangioma with some small cells in the stroma. There was also an epithelial inclusion cyst in the specimen. The present condition of the patient is not known.

CASE 6. Age, 15 years. This patient had a cavernous lymphangioma of the conjunctiva. A hemorrhagic edematous swelling of the semilunar fold of the left eye had first appeared when the patient was about three and had slowly become more prominent until age 15 at which time an excision was undertaken. The prominent clinical feature was the presence of recurrent hemorrhages in the edematous swelling. Microscopic examination of the excised tissue showed a cavernous lymphangioma of the conjunctiva with lymphocytic infiltration aggregated into follicles.

CASE 7. Age, adult. This patient noted a lesion of the bulbar conjunctiva temporally in the left eye. A similar lesion had been present on the right eye nine years before. The fate of the lesion on the right eye was not known. The excision of the lesion of the left bulbar conjunctiva revealed a cavernous lymphangioma together with lymphangiectasis.

CASE 8. Age, 7 months. The patient had exhibited since shortly after birth a lesion of the conjunctiva and cornea of the left eye which was thought clinically to be a dermolipoma. Microscopic examination of the excised tissue revealed a limbal epidermoid together with a cavernous lymphangioma.

CASE 9. Age, 23 years. This patient had a cavernous lymphangioma of the bulbar conjunctiva (Figure 3). A cyst of the bulbar conjunctiva of the right eye had been noticed ten years previously. On several occasions an attempt had been made to aspirate the cyst with a needle, but each time it had filled with blood and the attempt had been unsuccessful. Accordingly, about ten years after onset, the lesion was excised *in toto*. Microscopic examination of the removed tissue showed a typical cavernous lymphangioma. The present condition of the patient is unknown.

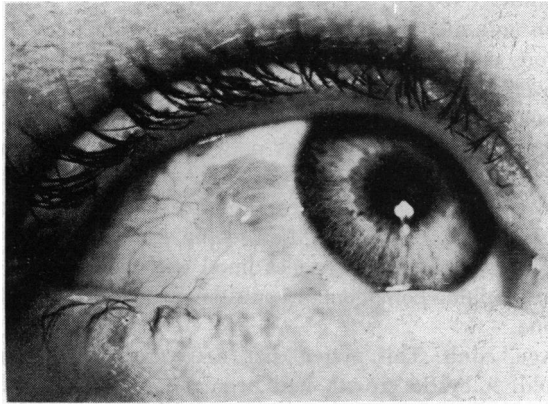


FIGURE 3. CASE 9

Lymphangioma of bulbar conjunctiva present ten years.

CASE 10. Age, 24 years. This patient had a cavernous lymphangioma of the inner canthus. The history was incomplete, but when the patient was about 21 years old, it had been noted that he had a mass involving the inner canthus. This was partially excised. Microscopic examination showed a cavernous lymphangioma with prominent stroma and with very few cells. Within three years the lesion recurred and again was excised. Examination of the specimen showed it to be similar to the first specimen except that the cavernous spaces were less evident and the stroma was more prominent. Scarring was apparent in the second specimen. The subsequent history of the patient is not known.

CASE 11. Age, 65 years. This patient had had an irritated eye for two weeks prior to surgery. A cyst was noted on the bulbar conjunctiva temporally and this was excised. Microscopic examination revealed a small cavernous lymphangioma. The present condition of the patient is unknown.

CASE 12. Age, 21 years. This patient had a cavernous lymphangioma of the conjunctiva. A mass had been present in the conjunctiva for an indefinite period of time and had been diagnosed as a hemangioma and removed three years before the surgery described here. The tumor recurred in the bulbar conjunctiva. Excision of the specimen and examination revealed a cavernous lymphangioma without other notable features. The present condition of the patient is unknown.

CASE 13. Age, 44 years. This patient had a cavernous lymphangioma of the bulbar conjunctiva. A cyst of the conjunctiva had been noted for two or three days. Excision and examination of this specimen revealed a cavernous lymphangioma with very few cells in the stroma. The present condition of the patient is unknown.

CASE 14. Age, 13 years. From the time the patient was five, it was noted that the inner quadrant of the conjunctiva of the left eye became red whenever the patient had a cold. A clinical diagnosis of hemangioma was made and a partial excision was performed. The condition recurred and two years later a partial excision again was performed. The condition recurred and one year later the third and final excision was performed. No recurrence in the five years since the third excision has been noted. Ten years after surgery the patient remains well and without recurrence. Pathologic examination of the tissue removed showed conjunctival epithelium with goblet cells and endothelial-lined cystic spaces. A light connective tissue stroma was present with a small lymphocytic element. The diagnosis from the microscopic examination was cavernous lymphangioma of the conjunctiva.

CASE 15. Age, 25 years. The early history was not obtained. When the patient was about 21, a mass was noted nasally in the bulbar conjunctiva of the left eye and a partial excision was carried out. Two years later this recurred and increased slowly and four years after the original excision a new excision was carried out. Microscopic examination of the tissue removed showed a cavernous lymphangioma of the bulbar conjunctiva with a moderate lymphocytic infiltration.

CASE 16. Age, unknown. Since the patient's childhood a mass had been present in the left upper fornix and this had slowly increased in size. Clinical examination showed a soft cystic avascular mass in the fornix when the upper lid was everted. This mass was excised at surgery and microscopic examination of the specimen showed a cavernous lymphangioma tending to the cystic type. The connective tissue stroma was sparse and there was very little lymphocytic element.

CASE 17. Age, 18 years. This patient had a cavernous lymphangioma of the bulbar conjunctiva. A mass had been noted in the bulbar conjunctiva for an indefinite period exceeding one year. Clinical examination showed an avascular mass approximately $6 \times 5 \times 3$ mm. Microscopic examination of the excised mass showed a cavernous lymphangioma with a slight infiltration of small lymphocytes.

CASE 18. Age, 20 years. This patient had a lump on the bulbar conjunctiva of the right eye which had been noted for five months previous to admission. A complete excision was carried out and microscopic examination of the tissue which was removed revealed a cavernous lymphangioma with small spaces and a delicate connective tissue stroma. There was a small cellular element.

CASE 19. Age, 28 years. This patient had a lymphangioma of the bulbar conjunctiva. For an indefinite period previous to admission but especially in the preceding three months a cyst had been present on the bulbar conjunctiva. This was excised completely and microscopic examination of the specimen revealed a cavernous lymphangioma with a small cellular element.

CASE 20. Age, 33 years. This patient had a cavernous lymphangioma with some features of ectasia. A lump had been noted on the bulbar conjunctiva for one month prior to surgery. The lump had been cauterized and had disappeared, but re-formed shortly thereafter. A complete excision and examination of this specimen revealed clear lymph spaces in a loose stroma. This was either cavernous lymphangioma or lymphangiectasia, or both.

CASE 21. Age, 44 years. This patient had a cavernous lymphangioma with some ectatic elements. An elevated lesion had begun laterally on the conjunctiva about one year prior to surgery. This was thought to be a pterygium and was completely excised. Microscopic examination of the tissue removed showed a cavernous lymphangioma with some lymphangiectatic elements.

CASE 22. A cyst had been present on the bulbar conjunctiva for eight months. A complete excision was carried out. Examination of the excised specimen revealed a lymphangioma.

LYMPHANGIOMAS OF THE LID

CASE 23. Age, 64 years. This patient had always had a lump on the outer aspect of the right upper lid. In the weeks before he came for examination, this lump had become larger. A clinical diagnosis of dermoid cyst was made. No X-ray pictures were taken. In the operating room an incision was made parallel to the brow and extending down to the lateral canthus. Blunt dissection revealed a cystic mass in the tissues and this was excised. During the dissection the cyst was opened and necrotic discharge was expressed. Microscopic examination of the tissues which were removed showed fibrous tissue, muscle, and orbital fat. Some areas of the fat showed localized lymphocytic collections. Other areas showed endothelial-lined spaces. The diagnosis was lymphangioma of lid and brow.

CASE 24. Age, 6 years. This patient was first admitted with a history of tissue twice previously being removed from the right side of his nose and lids since the age of one (Figure 4). The clinical diagnosis was dermoid cyst. Following the second removal the size of the cyst increased to maximum again within one year. A firm loosely anchored mass approximately $10 \times 8 \times 5$ mm. was found on the right side of the nose and lids. The tumor was



FIGURE 4. CASE 24

Lymphangioma of right side of nose and both lids present since age one and twice previously partially excised.

excised and was found to be a fibrous mass which was grossly a fibroma. A small fossa was observed in the nasal side of the orbit and nose where the mass had rested. Microscopic examination of the tissue showed it to be a lymphangioma.

CASE 25. Age, 8 years. This patient had a lymphangioma of the conjunctiva and lids (Figure 5). At the patient's birth, the left upper lid was noted to be swollen and there was a slow increase in the size of the lid until the patient was eight at which time a partial excision of the lesion was undertaken. Microscopic examination of the tissue removed showed cavernous endothelial-lined spaces with a moderate connective tissue stroma and with prominent blood vessels in the septa of the connective tissue. A prominent lymphocytic infiltration was present with follicles noted at one place. The present condition of the patient is unknown.

CASE 26. Age, adult. This patient had a cavernous lymphangioma of the right upper lid. A nodule had been noticed in the right upper lid six months prior to excision. The nodule had slowly grown larger and was excised for this reason. Microscopic examination of the tissue removed showed a cavernous lymphangioma with small, fairly uniform spaces and with a sparse cellular element.

CASE 27. Age, 8 years. This patient had a lymphangioma of the lid. Tissue had first been noticed prolapsed from the upper fornix when the patient was about two years old. A biopsy of the tissue was done, but the results are not known. When the patient was eight, an excision of the prolapsed tissue was



FIGURE 5. CASE 25

Lymphangioma of left side of nose and medial portion of each lid present since birth and slowly increasing in size.

undertaken and microscopic examination of the specimen removed showed a cavernous lymphangioma with large spaces and a delicate stroma. The cellular element was prominent.

CASE 28. Age, 1 year. This patient had a cystic lymphangioma of the conjunctiva and lids. A tumor involving the lids and conjunctiva on the left especially on the nasal side had been present shortly after birth. This swelling had become greater on straining and the conjunctiva protruded through the palpebral fissure. An incomplete removal of the lesion at surgery and examination of the specimen revealed a cavernous lymphangioma with large cystic spaces and some large blood vessels in the connective tissue septa. The cellular element was very small. The present condition of the patient is not known, and photographs are not available.

CASE 29. Age, 15 years. This patient had a cavernous lymphangioma of the left upper lid. A painless swelling of the left upper lid had been present since birth. There had been a gradual enlargement of the swelling, which was most prominent in the year before excision. The patient was given 400 roentgens of irradiation with about one-third decrease in the size of the lesion, and excision was then carried out. Microscopic examination of the removed tissue showed a cavernous lymphangioma with collapsed spaces and a prominent stroma. The cellular element was very sparse. The present condition of the patient is not known.

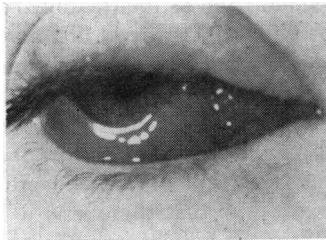
CASE 30. Age, 9 months. This patient had a cavernous lymphangioma of the right upper lid. He had had since birth an enlarged right upper lid which had increased in size tremendously following recent injury. The right

eye was closed due to swelling. Surgery was undertaken and a tumor was located extending from the skin down to periosteum. Microscopic examination of the tissue removed revealed a cavernous lymphangioma with tortuous thickened blood vessels and with a very sparse cellular element. The present condition of the patient is not known.

CASE 31. Age, approximately 12 years. This patient had been born with a congenital lymphangioma of the right side of the face and had had numerous episodes of cellulitis connected with it (Figure 6). Many of these episodes of cellulitis had followed injections of sclerosing solutions. Some of them had followed pharyngeal infection. The eye had not been noted to exhibit lymphangioma until two months before examination when it had become red and swollen following a cellulitis of the face. The facial cellulitis had cleared, but the eye had not. When the patient was seen in the eye clinic, she had a right exotropia, a full upper lid, a depressed inner canthus, and an angiomatous red bulbar conjunctiva. Tissue was removed for



(a)



(b)

FIGURE 6. CASE 31

- (a) Lymphangioma of right side of face since birth; many episodes of cellulitis.
(b) Close-up of same patient showing conjunctival lymphangioma which became apparent only in adolescence following cellulitis of face.

microscopic examination and revealed a cavernous lymphangioma. The present condition of the patient is not known.

CASE 32. Age, 3 years. The right upper lid of this patient had been swollen from birth. An operation for removal of a growth of the conjunctiva of the right eye had been performed when the patient was one year old and again when he was two. When seen at age three, he showed a pale elevation of the semicircular fold and lower fornix with dots of hemorrhage. Tissue was removed for examination and showed lymphangioma. Follow-up information is not available.

CASE 33. Age, 12 years. A swelling of the right lower lid and cheek had been present since birth. This had become ecchymotic from time to time. A firm bluish subcutaneous mass measuring 2.5×3.5 cm. could be made out. The mass was excised and on microscopic examination showed lymphangioma.

LYMPHANGIOMAS OF THE ORBIT

CASE 34. Age, 23 years. At the time of her first visit, this patient reported that she had undergone a mastoidectomy when she was five years old, that a growth had been removed from the roof of her mouth at the same age, and that she had had a swelling in her neck at the age of ten. Also, beginning about age 13, she had become aware of a small red swelling at the inner canthus of the right eye. She received several X-ray treatments and had several operative procedures on the right side of the face and neck but none on the eye. The X rays were directed to the temple region rather than to the eye directly. The patient's main complaint was of bleeding from the ocular portion of her tumor. Examination showed the right fissure to be smaller than the left and there was a palpable mass in the upper and lower lids with a large, fleshy, reddish brown mass covering the globe nasally (Figure 7). Palpable tumor tissue, communicating above and below with the lid masses, was felt in the temple all the way back to the ear. Diagnosis, made on previous plastic surgery specimens, was lymphangiohemangioma. Sclerosing solutions had been advised as the treatment of choice. Vision was 20/50 in the right and 20/30 in the left eye at first admission at which time the right globe was displaced 20 mm. down and 15 mm. laterally compared to the position of the left globe. X-ray pictures showed the left orbit to be normal and the right to be increased in all dimensions. The bone of the upper nasal quadrant of the right orbit also showed increased bone density and the adjacent frontal sinus was clouded. There were no dehiscences. The opinion of the roentgenologist was that there was a long-standing tumor, probably a hemangioma. When the patient was 27, a partial excision of this tumor mass from the lids, orbit, and temple was undertaken. Microscopic examination of the tissue which was removed resulted in a diagnosis of lymphangioma. Another pathologist studied the sections and gave a diagnosis of hemangiopericytoma. A third pathologist examined the specimens

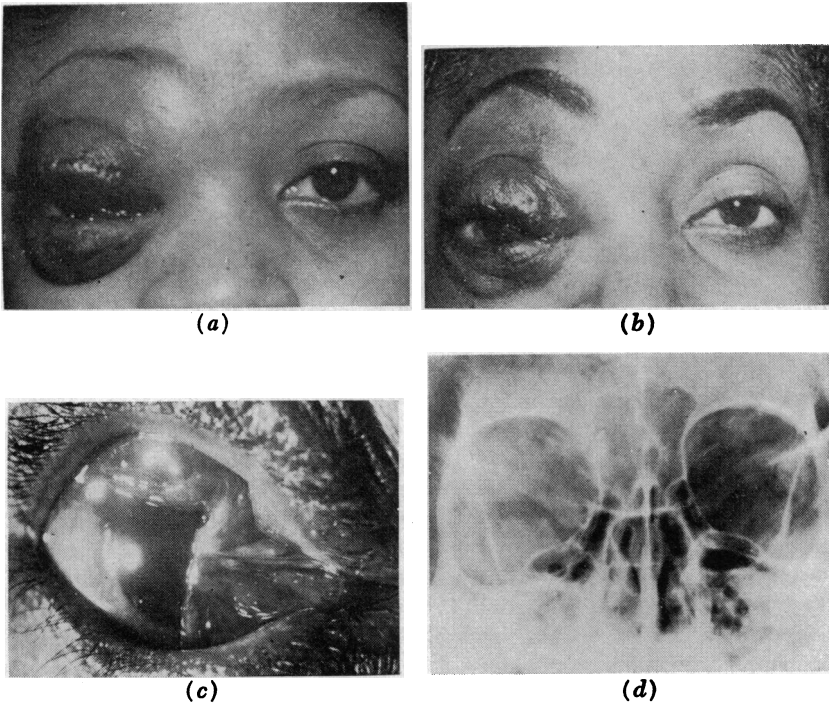


FIGURE 7. CASE 34

(a) Lymphangioma of the face, lids and orbit ten years after onset but before treatment; (b) appearance after surgery and irradiation; (c) close-up of the conjunctiva; (d) X-ray picture showing enlarged orbit.

and gave the diagnosis of "no pericytoma but hemangioma." A fourth pathologist, after review of all previous opinions, stated: "I think this is lymphatic rather than blood vascular; therefore, this is a cavernous cystic lymphangioma. The presence of follicular lymphoid deposits such as are seen in *hygroma cysticum* reinforces the impression of the lesion being lymphogeneous. Naturally it is benign." One year later an addition partial surgical removal was undertaken. On the third admission the patient showed an exophthalmos of 8 mm. on the right with minor improvement from previous surgery. Partial excision and plastic repair of the tumor tissue were again undertaken. The pathologist who examined the specimen microscopically, however, reverted to a diagnosis of hemangioma. At age 31 the patient again was admitted, the tumor having increased in size in the previous few months. Further excision and plastic repair, largely of the subconjunctival tumor mass of the right eye and orbit, were carried out. At the present time the most pressing problem is the rapidly re-establishing

pseudopterygium of the conjunctiva of the globe nasally on the right. This threatens to compromise the cornea and also threatens to extend between the lids. More surgery to attempt a removal of this is contemplated.

CASE 35. Age, 8 years. At the age of one, this patient injured his left eye by falling on the sharp end of a pencil which penetrated the upper lid. There was no damage to the globe. When he was two, a marked proptosis developed and at age two and two-thirds it began to recede spontaneously. X-ray pictures of the skull and orbits revealed nothing. When the patient was six, his eye again proptosed but receded spontaneously. At age eight, approximately three months before the present admission, the proptosis again developed during the course of a cold. Coincidentally, a left convergent squint developed. The ear, nose, and throat work-up was negative. The vision was 20/15 right and 20/70 left. The exophthalmometer showed 5 mm. of left proptosis. No tumor mass was felt. Visual fields were normal. Further X-ray pictures showed the left orbit to be larger than the right in all dimensions, but there were no dehiscences. The patient was taken to the operating room and through a Kronlein approach the orbit was opened. A tumor mass was palpated within the muscle cone and was exposed. It turned out to be soft, compressible, purplish blue in color, with a smooth capsule. During blunt dissection the capsule was ruptured and the mass collapsed. The type of fluid present could not be identified because of profuse bleeding from surrounding tissues. As much of the wall of the cyst as possible was removed. Microscopic examination of the tissue which was removed showed it to be a portion of the wall of a lymphangiomatous cyst possibly associated with an old hematoma. The wall was fibrous tissue without any recognizable

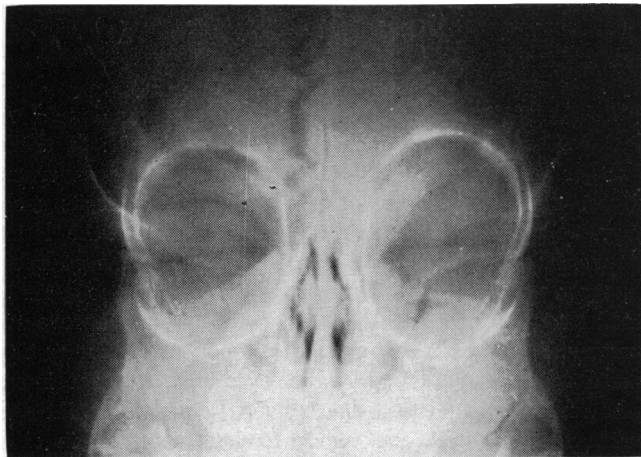


FIGURE 8. CASE 36

X-ray picture of a six-month old patient with lymphangioma of the orbit showing enlargement on the left.

lining cells. It was felt that the endothelial lining had been lost adventitiously.

CASE 36. Age, 6 months. This patient was first seen at age six months with a history of swelling of both of the lids of the left eye noted in the first week of life. This had been diagnosed as inflammatory originally, but persisted in spite of treatment. Between the ages of five and six months the swelling became considerably greater. There were no birthmarks elsewhere on the body. Clinical examination showed no exophthalmos, but there was a redness of the left side of the nose, left brow, lids, left cheek, and left side of the mouth. No mass was palpable and there was no obvious exophthalmos. Chemosis of the lower bulbar conjunctiva and palpebral conjunctiva with moderate injection were present. The caruncle was enlarged. A diagnosis of possible hemangioma of the left orbit was made. After consultation the diagnosis was amended to lymphangioma of the left orbit and face. X-ray pictures of the skull and orbits showed the left orbit to be larger in all dimensions than the right (Figure 8). There were no dehiscences of bone. The impression of the radiologist was that there was a congenital deformity involving the left orbit. A biopsy of the conjunctiva was undertaken and microscopic examination revealed a lymphangioma.

CASE 37. Age, 8 years. Five months previous to admission, following an attack of measles, the right eye of this patient had begun to protrude. The protrusion had increased slowly and two months prior to admission the exophthalmos on the right had been found to be 6 mm. The vision was unaffected. There was no bruit. Examination showed that the right eye protruded forward, down, and slightly to the side. Exophthalmos at the time of examination was 4 mm. The impression was that there was a retrobulbar tumor of unknown nature. X-ray pictures of the skull and orbits showed them to be comparable in size and shape. The patient was taken to the operating room and through a Kronlein approach the right orbit was explored. A tumor mass was felt above the optic nerve and a dark blue cystic tumor was brought into view. During attempted dissection the cyst broke and old blood was evacuated. The entire cyst apparently was removed from the orbit. The bone flap was repositioned and the soft tissues were closed. The specimen which was removed measured $19 \times 12 \times 11$ mm. Microscopic examination showed a multilocular cyst with an indistinct, thin, endothelial-lined surface. The wall of the cyst contained collections of lymphocytes with follicle formation. Some of the cells contained pigment which was thought to be the result of old blood. The diagnosis was lymphangioma of the orbit. The patient has remained free of symptoms since surgery.

CASE 38. Age, 15 months. Nine days prior to admission this patient had experienced a rapidly developing proptosis of the right eye together with a temperature of 99.4° . The temperature had subsided in one day. Examination revealed a firm proptosis of the globe, reddish in color, with no fluctuation (Figure 9). The proptosis measured about 10 mm. X-ray pictures of the



FIGURE 9. CASE 38
Orbital lymphangioma with hemorrhage and acute inflammation of nine days' duration.

orbits showed the right to be a trifle larger in all dimensions than the left. There were no dehiscences of the bone. The right orbit was explored through conjunctiva and a large multiloculated bluish cyst was encountered which extended over the upper one-half of the globe. The cyst was entered and dark chocolate-colored fluid escaped. The dissection of the cyst was carried posteriorly almost to the apex of the orbit. The entire cyst wall was not removed, but several representative specimens were. Microscopic examination of the tumor which was removed showed numerous large blood vessels in the specimen, some having thick fibrous walls. There were many small capillaries with evidence of old and recent hemorrhage. The diagnosis was angioma of orbit and hematoma of orbit. Re-examination of the specimen showed endothelial-lined spaces with clear fluid content. This appeared to be an orbital lymphangioma.

CASE 39. Age, 13 years. The patient was admitted with a history of a skin eruption, thought to be hives, which had occurred one month previously. Within a few days both eyes puffed up and a blood vessel ruptured on the right eye. The local eye doctor found a fluctuating mass, which was aspirated and dark old blood obtained. The examination showed a divergent right eye with dilatation of the anterior ciliary veins over the lower half of the limbus; 5 mm. of right exophthalmos were present (Figure 10). The vision was 20/25 right and 20/20 left. X-ray pictures showed increased orbital dimensions on the right together with increased soft tissue density. There were no dehiscences of bone. The provisional diagnosis was hemangioma of the right orbit and the patient was taken to the operating room where the orbit was explored through a Kronlein approach. A tumor mass was identified below the globe in the inferior part of the orbit and an attempt was made to dissect it out. During this attempt the capsule of the mass was broken and a large amount of dark old blood was evacuated. The remainder of the capsule was excised, the bone flap was replaced, and the tissues closed. The specimen consisted of large multilocular cysts without

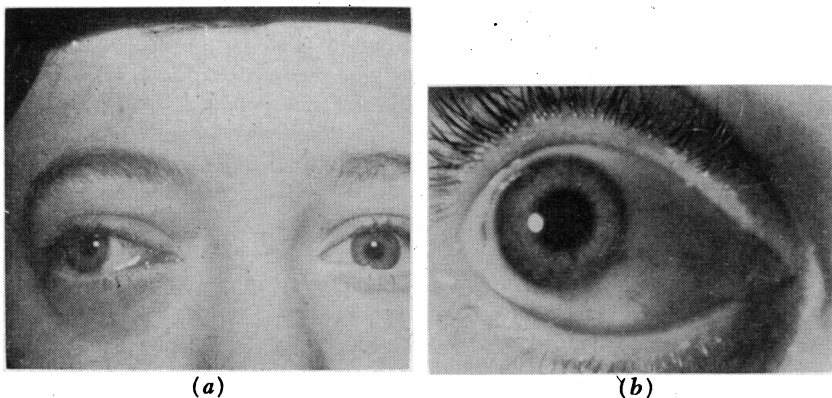


FIGURE 10. CASE 39

(a) Orbital lymphangioma with hemorrhage and increased proptosis; (b) close-up view showing remains of ecchymosis in the conjunctiva.

the usual well-formed endothelial lining. The lining cells were flattened and the walls of the cyst were irregular in thickness. The initial pathologic diagnosis was hematoma of orbit. Further examination indicated this to be a hemorrhage into a cystic lymphangioma of the orbit.

CASE 40. Age, 17 years. The first hospital admission of this patient occurred when she was 17, at which time she gave a history of having had a bloodshot right eye since age six. Beginning when she was about ten, her right eye had become prominent. A diagnosis of abscess behind the eye was made and an exploratory operation was carried out through the medial portion of the right upper lid with a partial ethmoidectomy. Several pieces of tissue were removed and showed a chronic inflammatory process. Following surgery the eye turned out. The vision at the time of the surgery was 20/30 in the right and 20/20 in the left. One year later the exophthalmos recurred and again a partial ethmoidectomy was performed with improvement. One year later the exophthalmos again recurred. This time no inflammatory process could be found. Iodides and mercury were given without results. The examination at the time of the patient's admission to the hospital in 1935, when she was 17, revealed 6 mm. of right exophthalmos with 45 diopters of exotropia. Her vision still corrected to 20/20 in each eye. Ecchymosis was present in the medial portion of the right upper lid. There was a weakness of the internal rectus and of the superior oblique. The examination was otherwise negative and a diagnosis of pseudotumor was made. X-ray films taken at this time showed marked asymmetry of the orbits. The left was normal and the right was strikingly enlarged in all dimensions. The impression of the radiologist was that this was a manifestation of long-standing increased intraorbital pressure. An orbital exploration

was undertaken through a lateral canthotomy incision. A soft lobular flattened mass was palpated along the lateral wall of the orbit extending into the apex. This was removed. The microscopic examination revealed lacrimal gland, striated muscle, and orbital fat, none of which was abnormal. Tissue removed at one of the previous operations was secured and showed chronic inflammation. Accordingly, a provisional diagnosis of pseudotumor was entered. The patient was readmitted two months later and an exploration was carried out through the bulging conjunctiva. A mass of subconjunctival fibrous tissue was removed from the floor of the orbit. Again a soft lobulated mass was felt in the orbit and a portion of it was removed. The specimen under the microscope showed fibrous tissue and fat with a lymphocytic infiltration. There were areas of old hemorrhage. Several irregular cavernous spaces with endothelial lining and clear fluid content were present. This was a lymphangoma of the orbit with hemorrhage into it. Three years after her original admission she was again admitted with proptosis of the right eye. This time the exophthalmos measured 5 mm. She was treated with pronylin without any change in appearance. (See Figure 11.)

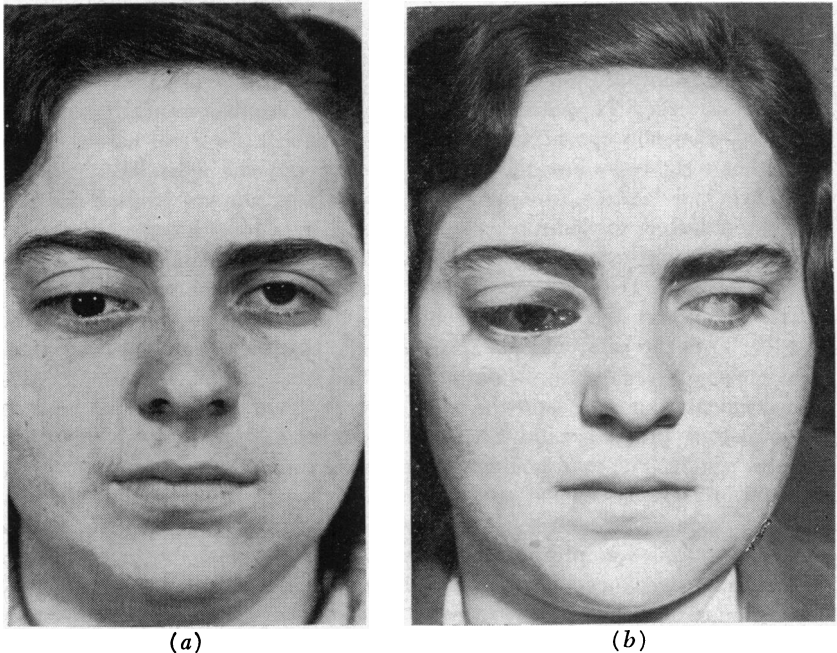


FIGURE 11

(a) Orbital lymphangioma with proptosis present constantly; (b) same case during increased proptosis and ecchymosis.

CASE 41. Age, 23 years. The right side of this patient's face had been swollen since she was four months old. When she was two and one-half years old, a biopsy specimen was taken and reported as lymphangioma. At age three the lesion was described as follows: "The upper surface of the right upper lid is the seat of a soft papillary-like lesion about 2×2 mm. The eyelid is swollen. The under surface of the lid shows a soft elevated lesion. The mucosal surface of the upper lid on the right side shows a soft papillary lesion." The patient experienced recurrent attacks of swelling and redness of the right cheek and eyelid at about monthly intervals. These attacks lasted about two to four days, sometimes with injection and discharge of pus from the eyes and the nose. Her temperature sometimes went to 104 during attacks. When she was eight, the lesion was described as a granular, bluish, mottled lesion of the right hard and soft palates typical of lymphangioma. At age 11 there was noted an area of ecchymosis at the right inner canthus which had been present for six weeks. Two days before the observation it suddenly became much larger. The mother of the patient associated the increase in the swelling with the onset of menstrual periods. At the inner angle of the right upper lid the raised red tissue was apparent with fullness of the orbit opposite this site. For the first time, 2 mm. of exophthalmos of the right eye were noted. Verruca-like lesions were present along the lid margin in the area previously diseased. Radiation treatment had been given and the patient showed a radiation cataract. At age 19, the patient had a marked subconjunctival hemorrhage and hemorrhage into the caruncle on the right. At age 22 a nodular area in the right cheek was noted and this was treated with radon seeds. At the time of admission, when she was 23, a biopsy of the subconjunctival tumor was undertaken in the operating room. Grossly the tumor was reddish and soft but somewhat fibrous. A partial dissection was carried out. During this dissection a blood cyst was encountered and ruptured, allowing the escape of some old dark brown blood. Microscopic examination of the tissue showed it to be characteristic lymphangioma with marked fibrous thickening of the tissue around the lymphatic vessels. X-ray pictures of the orbit showed no disturbance in size or shape. The patient was last seen at age 28 at which time there was no change in the appearance of the eye. Correspondence with the patient, now age 36, indicates that there still has been no change in the appearance of the eye.

CASE 42. Age, 42 years. When first seen, this patient gave a history of having had a "tumor of the right eye" at age 11 for which the eye was removed. At age 35, the patient's empty socket began to swell whenever she had a cold. The swelling would come and go and some pain and discharge from the socket occurred at the time of the swelling. Examination showed the entire region around the eye to be swollen. There was a hard round mass at the inner upper angle of the orbit and a similar but smaller firm mass was present in the lower portion of the orbit. The socket was filled with a

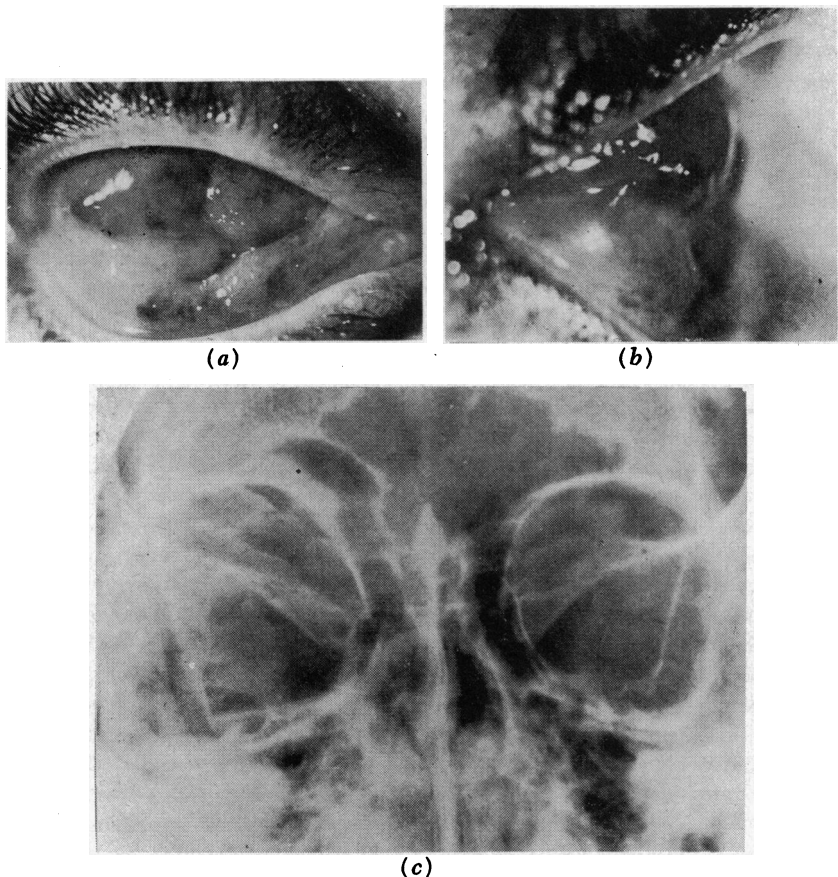


FIGURE 12. CASE 42

(a) Conjunctival sac showing lymphangioma which became apparent after eye had been removed for tumor; (b) side view; (c) enlargement of the orbit due to orbital lymphangioma.

soft, red, bloody tumor mass, irregular in shape (Figure 12 *a, b*). Provisional diagnosis was recurrence of orbital tumor, and the patient was admitted for surgery. X-ray examination of the orbits showed them to be quite asymmetric (Figure 12 *c*). The right orbit was much larger in all dimensions than the left. The bone of the lateral wall was thickened but without increased porosity. The sphenoid ridge on the right also appeared denser than on the left. A biopsy of the orbital tissue was carried out through conjunctiva and the specimen submitted to microscopic examination. This was reported as normal tissue. At age 41, the patient again presented her-

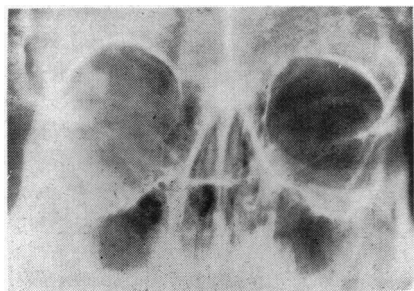
self saying that the right eye was painful and swollen and occasionally subject to bleeding. The examination was similar to that previously noted. A repetition of the X-ray examination was carried out and the pictures were read as showing a congenital deformity of the right orbit, possibly associated with neurofibromatosis. The patient was taken to the operating room for an excision of the orbital tumor. The conjunctiva could not be dissected off the underlying tissue. Accordingly, conjunctiva, soft tissues, and the tumor mass in the inferior portion of the orbit were all excised. Microscopic examination of the new tissue indicated the following: "The sections show a complex tumor made up of vessels; some are capillary, some show cavernous dilatation, others have smooth muscle in their walls and resemble atypical veins, and some seem to be lymphatics. However, these vessels are diffusely scattered throughout the orbital tissues and some of them are surrounded by fibroblastic proliferation. This is a nevoid type of growth coming from embryonally segregated vascular mesenchyme. Hemangiolympangioma of orbit." The patient was admitted at age 42 to a different service for excision of a tumor of the nasal cavity which was read as cavernous hemangioma. The patient was last seen at age 53, complaining of bloody tears from the right eye. The socket showed some mucopurulent discharge, conjunctival erosions, and serosanguinous discharge.

CASE 43. Age, 7 years. A swelling of the right upper lid had been noted for about two years. X-ray pictures showed the right orbit to be larger in all dimensions than the left (Figure 13). There were no dehiscences of bone. The impression was that there was an intraorbital tumor, nature undetermined. The patient was taken to the operating room where the orbit was approached from the front through a lid incision which was carried down to a dark thin-walled cyst. The cyst was opened and a copious watery brown fluid escaped. The interior of the cyst was explored digitally and found to ramify from the roof to the floor of the orbit on the medial side of the nerve. A considerable portion of the wall which presented was removed for microscopic study. A drain was left in place and the tissues were closed. Microscopic examination of the tumor which was removed led to a provisional diagnosis of hemangioma of the orbit, capillary type, but further study showed it to be typically lymphangiomatous with a large lymph-follicle element.

CASE 44. Age, 9 years. This patient's parents noticed a small lump at the inner canthus of the right eye shortly after his birth. This was thought to be a cyst. When the patient was about three, it was concluded that this lump was growing and should be excised. An intraorbital cyst connecting with this mass was removed when he was three and a half. It was thought to be a clean excision. A definite diagnosis was not made, but the tissue was said to be benign. Following the surgery, the right upper lid exhibited ptosis. Several times over the two years prior to the present admission, hemorrhages occurred with the lids and globe becoming black and blue. The



(a)



(b)

FIGURE 13. CASE 43

(a) Exophthalmos and presenting tumor due to blood cyst in an orbital cystic lymphangioma; (b) X-ray picture showing orbital enlargement.

patient's nose was always stuffy on the affected side. The swelling varied from day to day and the growth recurred in the upper lid (Figure 14 *a, b*). Just prior to admission, a diagnosis of angioma was made and surgery was not advised. Vision was found to be 20/40 in the right and 20/25 in the left; 5 mm. exophthalmos were present on the right and a smooth, rounded mass was palpable in the upper inner aspect of the orbit. The inner canthus and nasal conjunctiva had a reddish discoloration. X-ray pictures of the orbits showed the right to be a trifle larger in all dimensions than the left (Figure 14 *c*). There were no dehiscences of bone. The impression of the radiologist was that this might be a case of neurofibromatosis. Several consultants saw the patient and a provisional diagnosis of lymphangioma was made. In the operating room the conjunctiva was incised over the right caruncle and the underlying tissue was dissected out. An incision was made through skin over the upper nasal orbital margin, the tumor

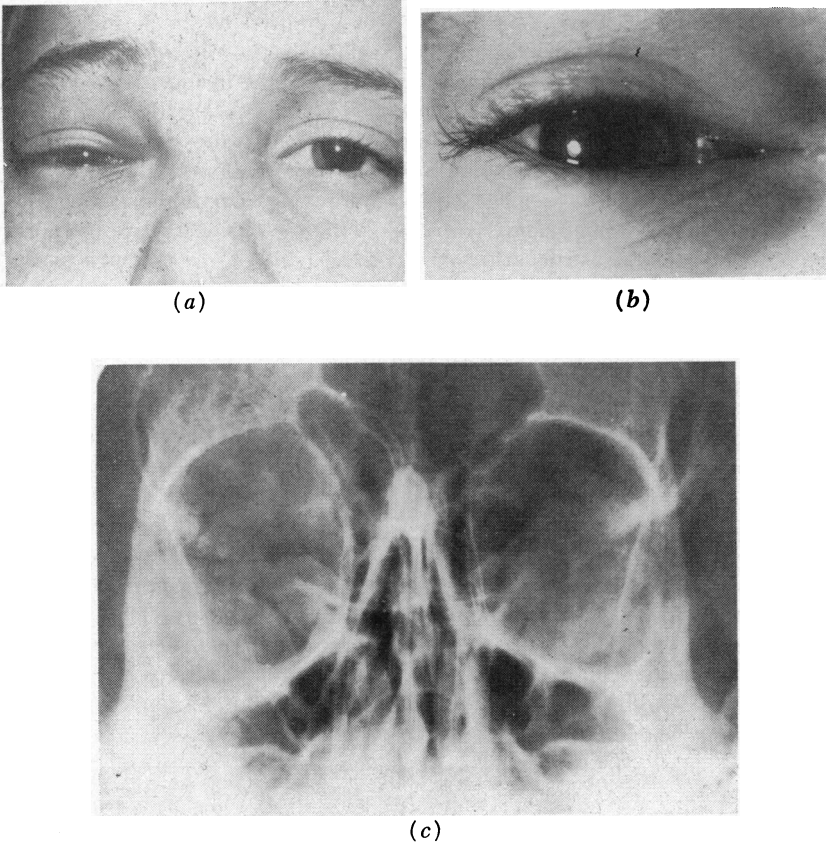


FIGURE 14. CASE 44

(a) Presenting tumor and slight proptosis right eye present since birth; (b) close-up showing swelling of lids, especially the upper lid medially, as well as conjunctiva swelling medially; (c) X-ray picture of the same patient showing enlargement of the orbit due to cystic lymphangioma.

mass was identified, and an attempt was made to excise it. It collapsed during the dissection, but a part of the wall of the tumor was removed for examination. Microscopic study revealed that the tissue consisted of large irregular spaces, lined with endothelium. Some of these spaces contained blood. Many thin-walled blood vessels were scattered throughout the specimen. In some areas there was a concentration of lymphocytes about the smaller vessels. The initial impression of the pathologist was that the tumor was hemangioma-cavernous, but further study indicated it to be a cavernous lymphangioma.

CASE 45. Age, 4 years. This patient was admitted to the hospital with a history of episodes of orbital and lid swelling in the right eye associated with ecchymosis and pain. These episodes lasted three to four days. A bruit had been heard at various times. Prior to admission a carotid clamp had been put in place, but the bruit disappeared and the clamp was not tightened. At the time of admission an acute exacerbation had been in evidence for two days but was spontaneously improving. A provisional diagnosis of orbital cystic lymphangioma was made (Figure 15). Pediatric consultation revealed no disease in other parts of the body. X-ray pictures of the skull and orbit showed the right to be slightly larger in all dimensions than the left. Carotid arteriography was performed and showed no abnormality. A radiotherapy consultation was obtained and it was agreed to give the patient some radiotherapy prior to surgery. The condition became entirely quiescent following this and the patient was discharged. Three days later the patient was readmitted with an acute exacerbation of the orbital swelling with tense skin, heat, pain, and a dusky red appearance. The patient was taken to the operating room and under general anesthesia a Kronlein approach to the orbit was carried out. No discrete orbital mass could be found, but there was a diffuse infiltration of the tissues. Several specimens were taken for microscopic examination. The lateral orbital wall was removed for decompression. Microscopic examination of the tissue which was removed proved it to be not entirely representative. One reading of the tissue resulted in a diagnosis of pseudotumor, lymphangioma, and hemangioma. Another reading led to a diagnosis of orbital granuloma, and a third reading was lymphangioma of the orbit.

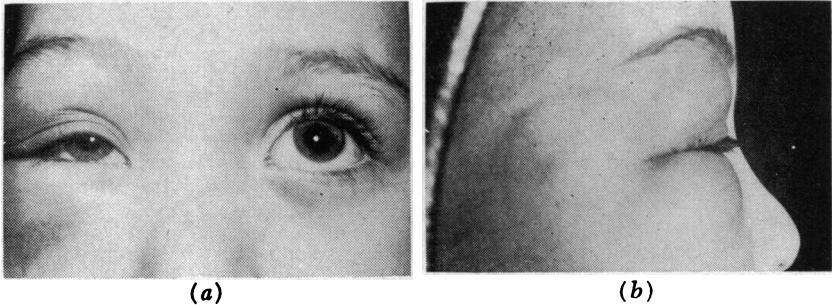


FIGURE 15. CASE 45

(a) Photograph showing acute inflammatory episode with proptosis and swelling of the lids due to lymphangioma in the orbit; (b) side view.

CASE 46. Age, 6 years. This child was first seen at age three with a history of having been hit in the left temple area with a bottle two years previously. Following this the patient developed a marked left exophthalmos. X-ray pictures taken at age three showed increased orbital dimensions on

the left side but no dehiscences of the bone. Surgery was recommended but refused at that time. The present admission at age six years was occasioned because of an increase in the exophthalmos of the left eye (Figure 16 *a*). The vision was still unaffected. Palpation revealed a soft fluctuant mass in the upper inner angle of the left orbit. A provisional diagnosis of hemangioma of the left orbit was made and X-ray pictures were taken. These demonstrated that the left orbit was still greater in capacity than previously. There were no dehiscences of bone (Figure 16 *b*). The impression of the roentgenologist was that there was a hemangioma of the left orbit. The patient was taken to the operating room and the orbit was explored through a Kronlein approach. A large ramifying purplish tumor, which appeared to be a hemangioma and which contained old blood, was excised in pieces, following which the bone flap was replaced and the soft tissues were closed. The tissues all showed dense scar tissue with heavy infiltration of lymphocytes and plasma cells. There were deposits of pigment which appeared to be old blood pigment. Fresh hemorrhage was present and a concentration of lymphocytes was seen around the large well-formed blood vessels. Several cystic endothelial-lined spaces were present. The pathologic diagnosis was lymphangioma of the orbit.

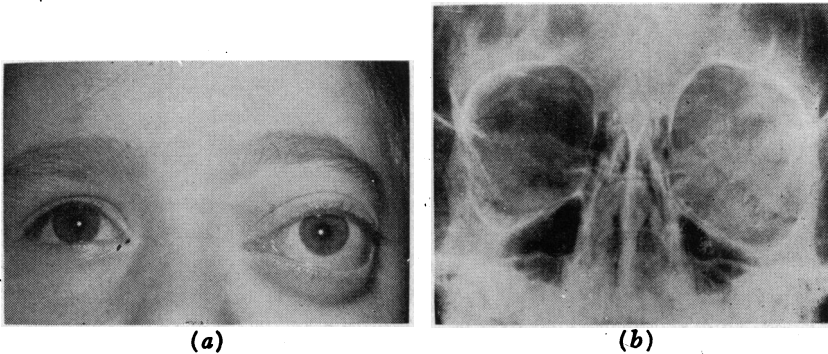


FIGURE 16. CASE 46

(*a*) Exophthalmos following trauma to the left eye. Surgery revealed a blood cyst in a lymphangioma. (*b*) X-ray picture showing orbital enlargement in this same patient.

CASE 47. Age, 19 years. At the time of admission to the hospital this patient gave a history of having had a lymphangioma of the left side of her face since she was two and a half years old. This had been infected at different times and several operations had been performed elsewhere. There was a severe proptosis of the left eye followed by loss of vision and a cyst formation on the eyeball (Figure 17). Partial removal had been tried, but the cystic area grew back. Aspiration was tried with no improvement. The

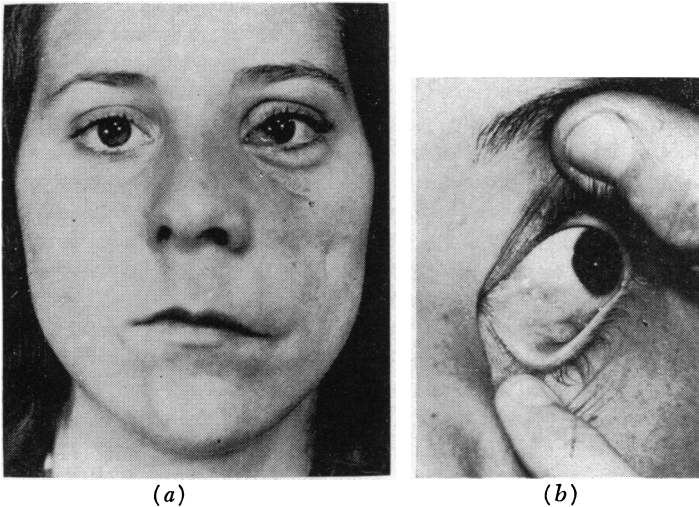


FIGURE 17. CASE 47

(a) Clinical appearance of lymphangioma of the face, lids, and orbit present since birth and subjected to surgery several times; (b) close-up view showing the conjunctival portion.

examination showed that there was no vision in the left eye. The inner canthus was displaced 4 mm. temporally, forward and down. Starting at the limbus and extending to the caruncle, there were large translucent cysts of the conjunctiva and smaller cysts of the fornix. X-ray pictures were taken and showed no difference in the orbits. There were several calcium concretions overlying the left antrum. In the operating room an excision of the subconjunctival cystic area was carried out. The microscopic examination of the specimen which was removed showed a dense fibrous tissue stroma with branching clear channels, lined with endothelium. Some of these contained blood. The diagnosis was lymphangioma. The present condition of the patient is not known.

CASE 48. Age, 5 years. Beginning when the patient was four, his right eye slowly became more prominent. At the time of admission 6 mm. of exophthalmos were measured. X-ray pictures of the orbits showed the right to have increased soft tissue density but no difference in orbital dimensions. The patient was taken to the operating room and a Kronlein operation was performed. A multiloculated bluish mass was immediately apparent behind the globe. In the process of dissection this was cut and dark degenerated blood escaped. Several pieces of tissue were taken for biopsy purposes. Microscopic examination of the tissue showed it to consist of many vascular spaces lined with endothelium, some of which contained blood. The diagnosis was lymphangioma of the orbit, cystic type.



FIGURE 18. CASE 49

Conjunctival manifestation of an orbital lymphangioma. This case was shown in Reese's textbook *Tumors of the Eye*.

CASE 49. Age, 11 years. This patient was first admitted to the hospital at age 11. He gave a history of having had a tumor of the left eye diagnosed as lymphangioma since age two. He had been admitted to other hospitals where he had repeated treatments with partial excision, sclerosing solutions, and radiotherapy. The history was of a swollen region around the left eye that extended from the bridge of the nose to the middle of the temple and below to the infraorbital ridge (Figure 18). There was moderate proptosis of the eye. Tearing had been present since birth. X-ray examination showed no bony defect in the orbit. The patient was taken to the operating room and an incision made into the conjunctiva of the upper nasal quadrant. The conjunctival tissue was grasped, dissected, and excised. Microscopic examination of this specimen showed lymphangiomatous channels lined with endothelium. These were partly obscured by a proliferation of blood capillaries which were thought to be secondary to the radiation treatment and not part of the lymphangiomatous process. The patient was last seen at age 13 when the possibility of exenteration of the orbit was raised. This patient has also been reported in Reese's book *Tumors of the Eye* (68).

CASE 50. Age, 45 years. The patient's history was that one year previously the right eye was noted to bulge. When admitted to the hospital, the patient was found to have 20/20 vision in both eyes with no appreciable correction necessary for the left and with one diopter of plus necessary for the right. Exophthalmometer reading showed 6 mm. of right exophthalmos (Figure 19). X-ray pictures showed the right orbit to be larger than the left and to have a defect in the frontal process of the zygoma. This was felt to represent invasion of the bone by an orbital tumor. Surgery was carried out and a blood cyst of the orbit was found. This was found in the floor and

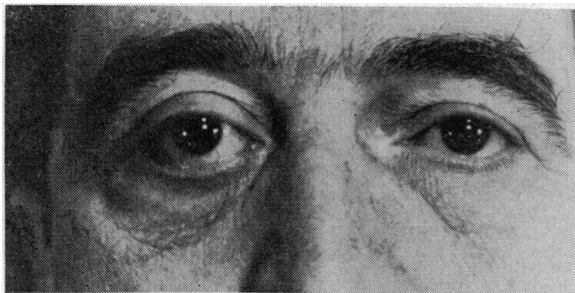


FIGURE 19. CASE 50

Right exophthalmos developing in an adult and found to be due to a blood cyst in a lymphangioma.

lateral portion of the orbit and extended about half way to the apex. The cyst was ruptured and brownish red mottled fluid escaped. Pieces of the orbital wall were removed for laboratory study. A defect was found in the bone which corresponded to the blood cyst. Microscopic examination of the specimen showed it to be the wall of a cyst with no epithelial elements noted. All layers showed an infiltration of lymphocytes with a few polyps. A layer of pigment-bearing cells just beneath the inner surface of the cyst wall was noted and this was thought to be hematogenous pigment. The pathologic diagnosis was blood cyst of the orbit. The specimen conforms to a lymphangioma cysticum in which the endothelial lining cells have been lost. This patient has previously been reported by Dr. John Wheeler as an orbital blood cyst (84).

CASE 51. Age, 10 months. The patient was first admitted at the age of ten months with a history of a soft tumor mass in the upper lid above the caruncle on the left side which appeared soft and compressible but became firm and engorged whenever the child cried (Figure 20). Injections into this tumor mass had been done elsewhere. A provisional diagnosis of hemangioma was made. X-ray examination revealed no changes from the normal. Aspiration was successful in removing only a few drops of tissue which were examined microscopically and found to be blood. At the age of two years the patient was readmitted and a similar examination was done. An excision of the orbital tumor was carried out. At operation it was found to be tightly connected to conjunctiva but to extend deeply. It was removed in a single piece and was a large, red, fleshy, compressible tumor. Microscopic examination of the specimen showed it to be made up of many vascular channels surrounded by hyperplastic endothelium. Some of the channels were filled with blood. A provisional diagnosis of hemangioma was made. The specimen conforms to the diagnosis of lymphangioma containing adventitious blood. The patient was seen again at age 12 and her vision was found to be 20/15 right and 20/200 left. She was last seen at age 16 at

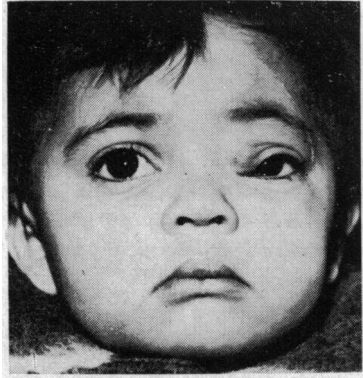


FIGURE 20. CASE 51

Lymphangioma of the orbit and left upper lid prior to surgery. The patient obtained a good cosmetic result.

which time there was no exophthalmos and no remaining sign of the old lymphangioma.

CASE 52. Age, 12 years. This patient had a cystic hygroma or lymphangioma of the orbit. The history was of a proptosis of the right eye with a slow increase which caused the patient to be taken for medical attention first at the age of seven and a half. Observation only was carried out. Five years later the proptosis amounted to 5 mm. (Figure 21). Surgery was undertaken and a specimen was removed. Microscopic examination of the tissue revealed a cystic type of cavernous lymphangioma of the orbit with a prominent lymphocytic element and lymphocytes aggregated into lymph follicles. The present condition of the patient is not known.

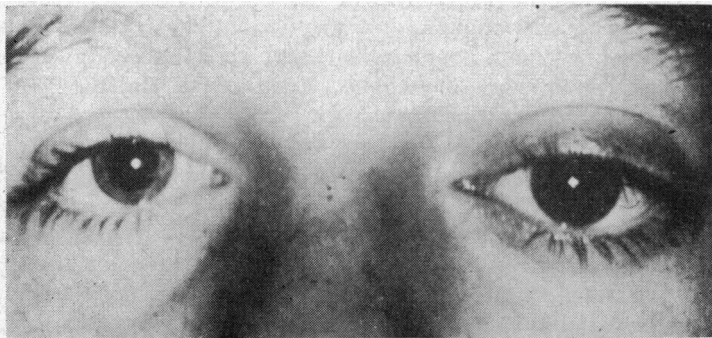


FIGURE 21. CASE 51

Slight exophthalmos on the left in a patient with orbital lymphangioma.

CASE 53. Age, 5 years. This patient had a lymphangioma of the orbit (Figure 22). The history was of progressive exophthalmos since birth. A biopsy had been taken at age five. The biopsy diagnosis was lymphangioma. The exophthalmos and disturbance of orbital contents were such that an exenteration was undertaken. The specimen was from the globe and orbital contents. Examination showed a lymphangioma of the orbit which apparently arose around the posterior ciliary arteries and which was marked by cavernous spaces with an endothelial lining and a very prominent round cell element. The lesion compressed the optic nerve. The present condition of the patient is unknown.

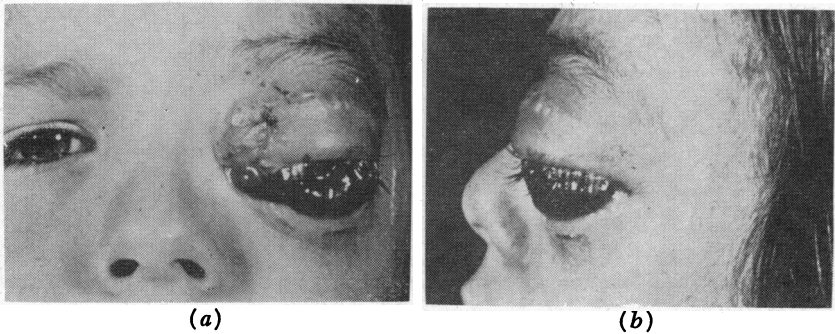


FIGURE 22. CASE 53

(a) Lymphangioma of orbit, conjunctiva, and lids present since birth; (b) side view of same patient. Progression led to exenteration.

CASE 54. Age, 2 years. This patient had a cavernous lymphangioma of the orbit. Since birth the globe on the affected side had been proptosed and the surrounding tissues had been edematous. A biopsy had been undertaken earlier, but the results are not known. The patient presented at the age of two a recurrent ecchymosis in the lids with swelling of the lids and proptosis of the globe. An excision of the orbital tissue was undertaken which was probably incomplete. Examination of this specimen showed a cystic variety of cavernous lymphangioma with very large spaces and many collections of lymphocytes. The stromal blood vessels and the connective tissue stroma were rather prominent.

CASE 55. Age, 19 years. This patient had a lymphangioma of the orbit. The left orbit since birth had contained a slightly proptosed globe with an upper lid which was slightly low. Excision was carried out in infancy, but the condition recurred. A partial excision was carried out when the patient was 13, but again there was a gradual recurrence so that at age 19 proptosis and ptosis of the upper lid were present. An excision, probably only partial, was carried out and examination of the tissue removed revealed a cavernous lymphangioma with a small cellular element and with prominent blood vessels in the connective tissue stroma.

CASE 56. Age, 5 years. This patient had a cavernous lymphangioma of the orbit with a cystic element. Since birth a pea-sized lump had been present in the upper inner angle of the right orbit and a gradual enlargement had taken place. A complete excision was carried out and the specimen when subjected to microscopic examination revealed a cavernous lymphangioma with a cystic element. There were large blood vessels in the connective tissue septae and the lymphocytic element was aggregated into follicles in some places.

CASE 57. Age, 11 years. This patient was first seen at the age of two years at which time he gave a history of congenital hemangioma involving the right upper lid nasally. He had received injections of sodium morrhuate beginning when he was about one year old and also CO₂ snow treatments to the skin. The original examination showed pigmentation of the skin of the right upper lid medially, thickening of the skin, and large dilated vessels filled with blood in the upper fornix (Figure 23 *a, b*). The medial canthus was displaced somewhat downward, and the globe was exotropic but not proptosed. It was felt that the mass extended posteriorly into the orbit. The original diagnosis was hemangioma of the right upper lid. The patient was admitted for plastic surgery, but this was deferred because it was felt that nothing was to be gained. Several months later the patient was again signed in and a plastic procedure designed to elevate the lid was carried out, but no excision of tumor tissue was done (Figure 23 *c*). X-ray pictures of the orbits showed the right to be greater in all dimensions than the left (Figure 23 *e*). There were no dehiscences. Three years later, when the patient was five, he was again sent in for a partial excision of this alleged hemangioma. At surgery the mass in the upper lid and orbit was found to be pulsating and to contain blood. A partial excision was carried out (Figure 23 *d*). The patient's recovery was uneventful although he reported that about once a month blood was on his pillow in the morning. Some further injections of sodium morrhuate were carried out. At age 11, the patient was again admitted for partial excision of the tumor mass and a plastic repair was carried out. (Figure 23 *f*). The lid was sacrificed in its medial 5 mm. and a dissection subconjunctivally of the tumor mass was also carried out. Bleeding was heavy and in some instances arterial. The tissues were scarred and the blood channels were large. Microscopic examination of the tissue removed at this time revealed it to be a typical lymphangioma of the cystic variety. The channels were large and irregular, contained an endothelial lining and contained lymphocyte aggregations in the surrounding tissue. The original lesion was either a lymphangioma or a combined lymphangioma and hemangioma, or a lymphangioma secondarily invaded by blood vessels, either as the result of trauma or as the result of sodium morrhuate injections given early in life. The subsequent course was felt to be out of keeping with any known hemangioma in that the progress was slow but inexorable and refractory to treatment.

Although the course may continue to be one of slow progression, the present situation is the most favorable, as far as cosmetic appearance and function goes, that the patient has known (Figure 23 g).

CASE 58. Age, 20 years. Since the patient was five, his right eye had protruded when he leaned forward. The exophthalmos was negligible when the patient was erect but 9 mm. when he was bending forward. X-ray pictures showed the right orbit to be larger than the left. Partial excision of an orbital cystic tumor was done. The pathology report was lymphangioma. Seven months later further surgery was performed without improvement. Following his last procedure the patient had a residual proptosis which increased when he bent forward and he felt that the condition was aggravated during a tour of parachute duty. Accordingly, in 1951, elsewhere, he underwent a transcranial approach to the orbit with excision of a portion of the tumor. Examination at age 28 showed the eyes to be essentially normal except that the right eye, which was the affected one, was less prominent than the left when the patient was in an erect position. When the patient bent over for about 30 sec., the right eye developed 8 mm. of exophthalmos which disappeared in one min. after the patient became erect again.

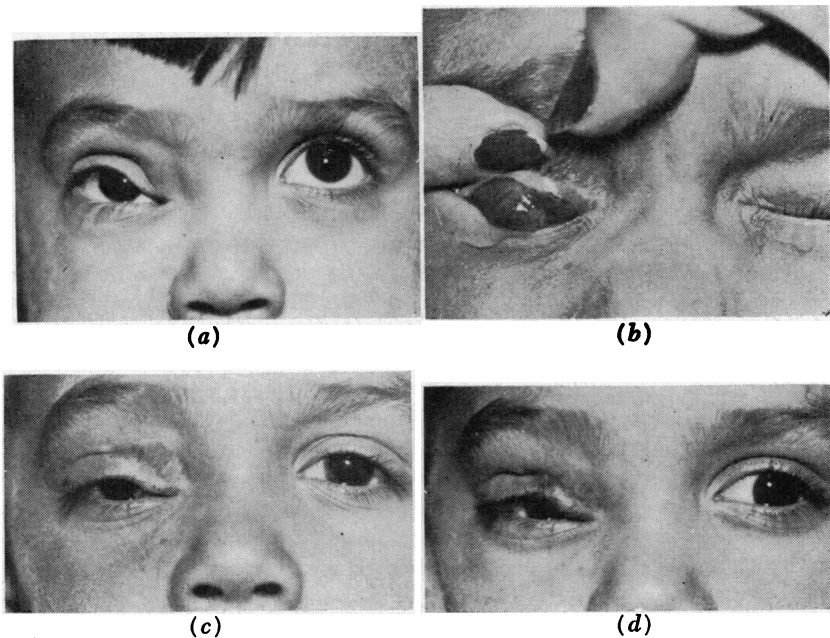
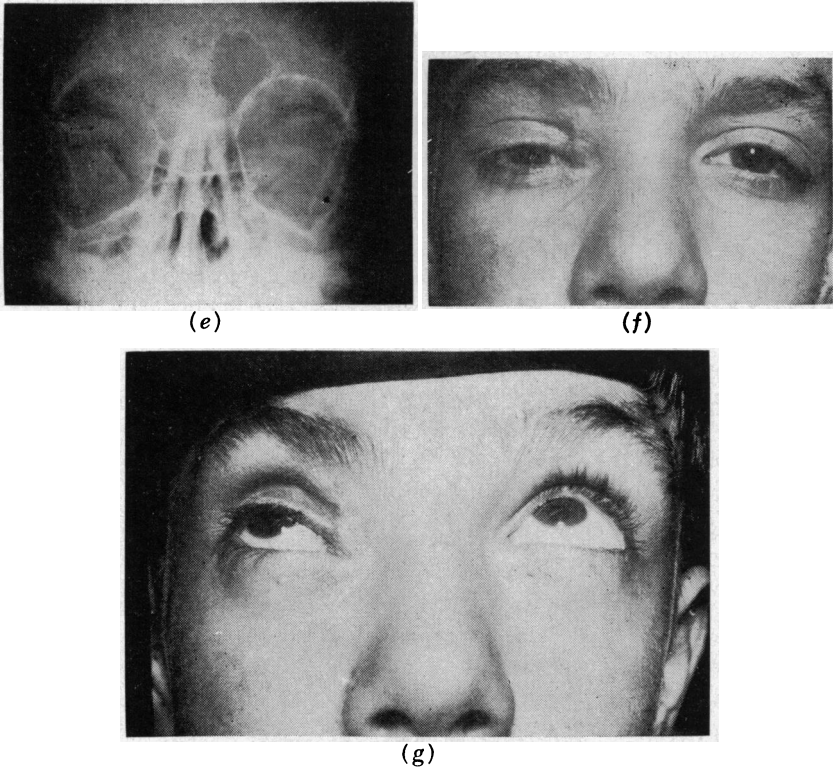


FIGURE 23. CASE 57

(a) Lymphangioma of orbit, upper lid, and fornix shown early in its development; (b) the upper lid everted to show tumor mass in fornix; (c) the same patient following sclerosing solutions and surgery; (d) following further surgery.

**FIGURE 23. CASE 57**

(e) X-ray picture of the orbits showing an increase in all dimensions on the right with an occluded adjacent frontal cell and change in the bone at the upper inner orbital angle; (f) the patient following recent partial excision and plastic repair; (g) second view taken recently to show that the ptosis is decreasing.

CASE 59. Age, 15 years. When the patient was four, excision of an orbital tumor had been carried out. The diagnosis was lymphangioma. The patient had undergone radiation with good results, but the tumor recurred. At examination, 4 mm. of left exophthalmos was present. A soft mass was felt in the orbit temporally. Fullness was present in the cheek. X-ray pictures showed a defect in the lateral wall at the sphenoid fissure. Additional treatment has not yet been carried out.

CASE 60. Age, 48 years. Ten years prior to admission this patient had a lymphangioma of the conjunctiva and orbit on the left which subsided when treated by injection. One year prior to admission there a recrudescence of the lesion which subsided without treatment. The present episode was three months in duration. Sometimes the swelling showed hemorrhage and

at other times not. A lump in the upper inner angle of the orbit had been present all his life. The mass had changed size from time to time. Examination showed a pedunculated red fleshy mass on the left semilunar fold and the fornix above and below. A rubbery mass in the orbit was palpable up and in. There was 1 mm. of exophthalmos. The clinical appearance at the last examination indicated hemangioma, but the diagnosis had been proved to be lymphangioma by an earlier biopsy.

CASE 61. Age, 29 years. The patient had increasing proptosis of the right eye for two years (Figure 24). Weakness of the superior rectus and the superior oblique muscles developed. X-ray pictures showed atrophy of the orbital roof. At operation an orbital blood cyst was drained and partially excised. The microscopic examination showed a fibrous tissue wall without lining cells. The initial diagnosis was orbital blood cyst, but further study showed the tissue to be compatible with a blood cyst in a cystic lymphangioma. This case was reported by Dr. John Wheeler (84) as an orbital blood cyst.

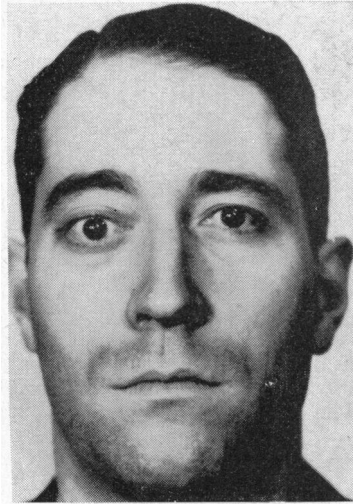


FIGURE 24. CASE 61

Right exophthalmos due to orbital blood cyst in a lymphangioma.

CASE 62. Age, 18 years. Since birth this patient has had a tumor in the medial half of the left brow. At age two a partial excision followed by radiotherapy was done. Facial deformity resulted. At age 16, intermittent bleeding began from a red swollen upper lid. At age 18, tumor masses were visible or palpable on the hard and soft palate, the cheek, the brow, the lids, the conjunctiva, and the lid margins (Figure 25 *a, b*). Exophthalmos

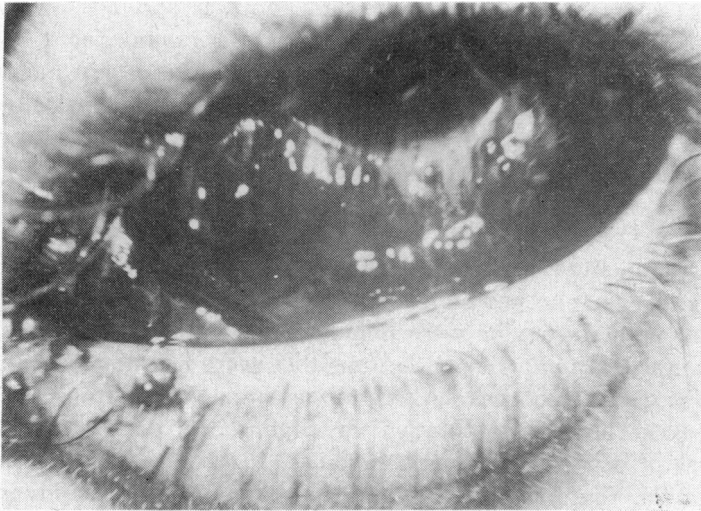
was present. The lid margin lesions were wartlike. X-ray pictures showed an enlarged orbit (Figure 25 *c*). A biopsy through the lower fornix revealed endothelial-lined spaces containing clear fluid and nests of small blood vessels. The diagnosis was lymphangioma with post-radiation blood vascularization.

ANALYSIS OF CASES

The 62 cases presented here were divided as follows: purely conjunctival lymphangiomas accounted for 22 of the cases or approxi-



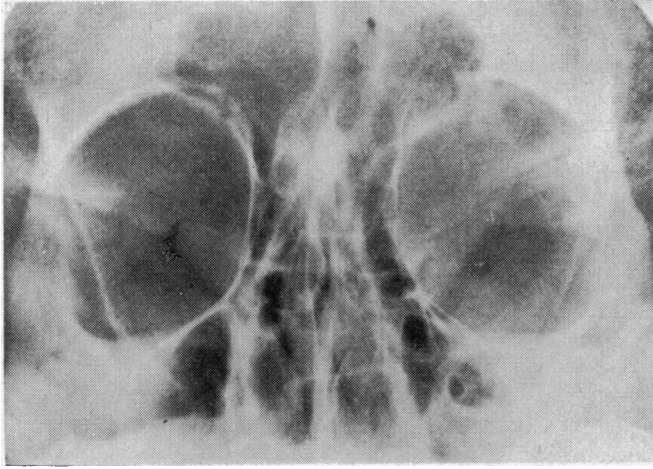
(a)



(b)

FIGURE 25. CASE 62

(a) Lymphangioma of orbit, conjunctiva, and lids with prominent hemorrhagic element; (b) wartlike tumors on the conjunctiva and lids.



(c)

FIGURE 25. CASE 62

(c) X-ray picture showing orbital enlargement.

mately 35 percent; those involving the lids numbered 11 cases or 18 percent. This included those in which the lids and conjunctiva were both involved and also those cases in which the face and lids were both involved. The largest group in the present series numbered 29 and comprised lymphangiomas affecting the orbit. This group amounted to 47 percent of the total. Included in it were lymphangiomas affecting not only the orbit but also the lids, the conjunctiva, and the face, in conjunction with orbital involvement.

CONJUNCTIVAL LYMPHANGIOMAS

The conjunctival lymphangiomas were noted to be present in patients varying in age from newborn to 65 years. The average age at which the conjunctival growths were noted was 25 years. The duration of the conjunctival tumors from the time they were first noticed until the time of excision, upon which this study is based, varied from two days to 12 years. The average duration was slightly over three years. Of the 22 cases of conjunctival lymphangiomas, 16 occurred on the bulbar conjunctiva. Four cases occurred on the semilunar fold and inner canthal region, one case was at the limbus, and one was in the upper fornix. The presenting symptom in 16 of the 22 cases was a discernible mass or lump. Five of the patients complained of a red eye, either recurrent or constantly present. One patient had recurrent hemorrhages

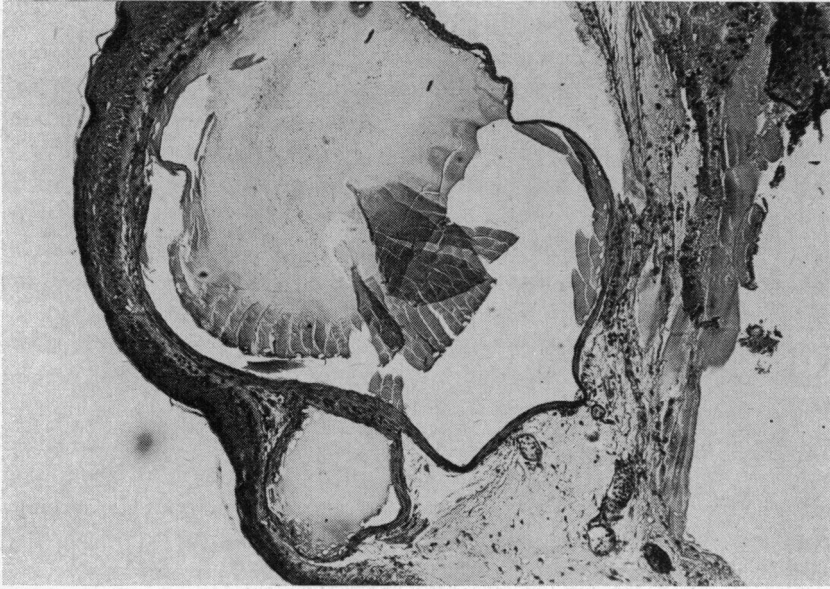


FIGURE 26. LYMPHANGIECTASIA

The simple dilation of a pre-existing channel is in contrast to the ramified spaces of true lymphangiomas. This section is not from any patient reported here.

into an elevated area. Although all of these patients were ultimately treated by excision, two had cautery preceding the final excision and one had unsuccessful aspirations attempted prior to the final excision (Case 9). One patient had a partial excision performed three times prior to the total excision. Two of the patients had partial excision performed three times prior to the total excision. Two of the patients had partial excision performed twice, and four had partial excisions performed once prior to the total excision. Examination of the tissues excised under the microscope revealed one lymph cyst together with an epithelial cyst. The remaining 21 specimens were all cavernous lymphangioma. Two of these lymphangiomas were associated with lymphangiectasia (Figure 26). Case 1 (Figure 1) had a pigmented nevus in association with the lymphangioma. One of the cavernous lymphangiomas tended toward the cystic type. One was associated with an epithelial cyst, and one was associated with an epidermoid of the limbus. Follow-up information was not available in most of the cases. The present status of 17 patients is not known. Five patients had no recurrence at the most recent period of observation.

LYMPHANGIOMAS OF THE LIDS

This group comprises 11 cases. The age of onset of eight of these was birth. One was first noticed at one year of age and another at two years of age. A single case did not become apparent until the patient was an adult. The average age of onset, therefore, was two years. The duration of the cases before treatment ranged from six months to 64 years. The average duration was approximately 13 years. The area most frequently involved was the upper lid. Eight of the 11 cases involved the upper lid, two involved the medial canthal area, and one involved the lower lid. The commonest symptom was a slowly increasing mass. Six of the patients had this history. Two of the patients had masses which from time to time exhibited ecchymosis. One patient had a mass which increased rapidly in size after injury. One patient exhibited a mass which increased in size on straining, and one other was not appreciated to have a tumor until it became more prominent after a cellulitis of an adjacent portion of the tumor. Six of the patients had complete excisions of the lymphangioma, but one of these had previously had two partial excisions and another had previously been treated by irradiation. Five patients were treated by partial excisions, three of these for the second or third time. One of the five had had sclerosing solutions injected into an adjacent facial lesion. X-ray examinations were not reported on any of these lid patients, but one individual was noted to have a fossa in the bone at the time of surgery. Microscopic examination of the tissue removed in all cases revealed cavernous lymphangiomas. Two of these had a prominent lymphocytic element with follicles being present in one. Another had large tortuous blood vessels in the lesion. Follow-up information on most of the patients was sparse. One patient was definitely cured, and one patient was not improved by surgery. The remainder were improved in the early follow-up period, but the final outcome was not recorded.

ORBITAL LYMPHANGIOMAS

The age of onset in this group of 29 patients averaged 6.2 years. Ten patients had the lymphangiomas at birth, 11 patients developed the disease in the period from birth to five years, and an additional five first showed the disease between six and 15 years of age. One adult aged 27, one aged 38, and one aged 44 first showed the lymphangioma at that time. The duration of symptoms from the time of onset until the treatment period upon which this study was based varied from nine days to 31 years, with the average being 8.3 years. Of these orbital lymphangiomas 13 were confined to the orbit. In addition to

orbital involvement, 11 of the patients had extension of the tumor to the lids or the conjunctiva. Five patients had lymphangiomas involving orbit, lids, conjunctiva, and face. Three of these five also had lymphangiomas of the palate. All of the patients had exophthalmos. Six of the 29 patients had an intermittent exophthalmos or an exophthalmos which varied in amount from time to time. Of these six, two had an increase in proptosis associated with coryza, one had an increase coincident with menses, one patient showed increased swelling associated with crying, and another patient demonstrated an increase when he bent forward. Among the exophthalmos cases were six associated with ecchymosis or hemorrhage. One of these six patients had an external hemorrhage from the conjunctiva from time to time. Four patients gave an antecedent cause for the development of their proptosis: one patient was normal until an attack of hives; another developed exophthalmos after a blow to the brow; a third showed ocular lymphangioma only after cellulitis in a facial lymphangioma; and a fourth developed a rapid exophthalmos subsequent to a high fever. The most common complication of orbital lymphangiomas was muscle imbalance. The five cases which exhibited this were distributed as follows: three showed an exotropia, one showed an esotropia, and one showed a weakness of the right superior rectus and the right superior oblique. The next most common complication was ptosis of the upper lid. Two patients exhibited this. An additional two patients had a concomitant sinusitis which may have represented lymphangioma involvement of the affected sinuses. One patient showed a cellulitis in a facial lymphangioma apparently continuous with the orbital lymphangioma, and one patient had a radiation cataract as the result of X-ray treatment of the orbital lymphangioma. X-ray pictures were available for study on 22 of the 29 cases of orbital lymphangioma. Of these, 16 showed an enlarged orbit on the affected side. Two of these 16 also showed sclerosis of the bone adjacent to the lesion, and an additional two showed a defect in the bone. Six patients showed no positive X-ray findings. All of these cases were interpreted as cavernous lymphangioma or the cystic variety of cavernous lymphangioma after a study of the sections. Three, in addition, had hemangiomas associated with lymphangioma. Eight of the patients had blood cysts in the cystic orbital lymphangioma spaces. Four patients had prominent blood vascular elements in the lymphangioma tissue. All of these last four had received either sclerosing solutions or radiation, or both, at some earlier period in their treatment. Two patients whose tumors were interpreted as lymphangioma of the orbit had lost the endothelial lining

of the lymphangioma spaces in the specimens examined. Follow-up information on the patients revealed three presumably cured, 15 improved, seven unimproved, and four whose present status is not known.

PATHOLOGY OF LYMPHANGIOMAS

The pathology of lymphangiomas as outlined in standard texts on pathology is applicable in general to the eye, but the peculiarities in this region deserve attention. Conjunctival lymphangiomas grossly were small lesions in this series and most of them were subject to complete extirpation. Many were so small as to escape detection until stasis produced a dilatation of adjacent lymph vessels and called attention to the lesion. These lesions often appeared to be clear conjunctival cysts. Microscopic examination of conjunctival lymphangiomas showed them to be composed of small closely packed endothelial-lined tubes lying in a very sparse stroma with a small or non-existent lymphocytic cell element. Some of the cases showed an adjacent endothelial-lined cyst, perhaps as large as the entire remainder of the lesion, which was lymphangiectatic. Case 22 (Figure 27) was clinically a solitary cyst.

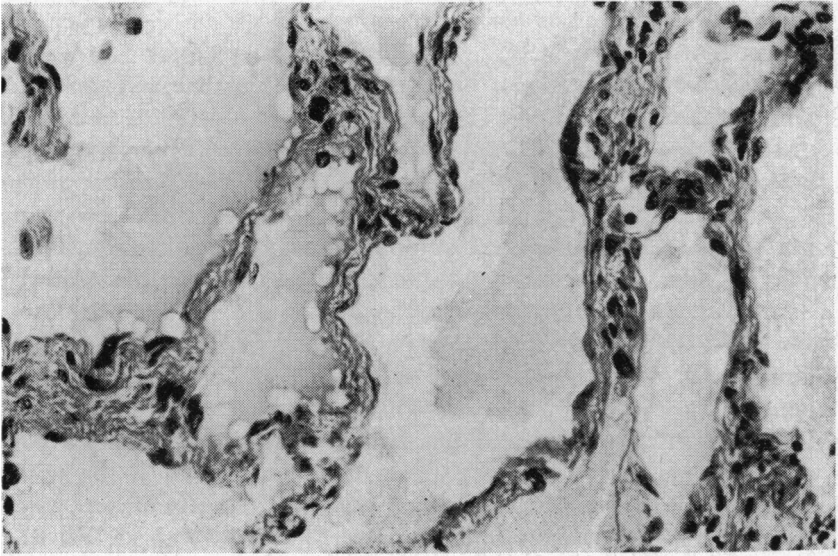


FIGURE 27. CASE 22

This cavernous lymphangioma of the conjunctiva demonstrates fairly uniform spaces, delicate septae, an endothelial lining, and albuminous fluid in the spaces.

The gross pathology of lymphangiomas involving the lids appeared less clear cut than that of the conjunctiva. The lesions were poorly demarcated. Accordingly, the gross specimens varied in size according to whether the specimen was a partial excision, a complete excision, or a complete excision with surrounding normal tissue. Most of the lid specimens were incomplete excisions done for purposes of establishing the diagnosis or in the course of a series of plastic surgical procedures. A striking feature in lid lymphangiomas was the variation in the amount of stroma. Some cases such as Case 34 (Figure 28) showed the stroma almost entirely replaced by endothelial-lined spaces. Others showed scattered spaces in a heavy stroma. The endothelial-lined spaces tended to vary in size with smaller spaces lying near the surface and larger spaces being more deeply placed. The higher power section from Case 34 (Figure 29) illustrates this. The lymphocytic element also varied from very little to a heavy amount aggregated into lymph follicles. Extensive lymphangiomas of the lids in which the conjunctiva was also involved frequently showed prominent large blood vessels. These may have been the result of repeated hemorrhages and vascularization of a portion of the lymphangioma, or of secondary

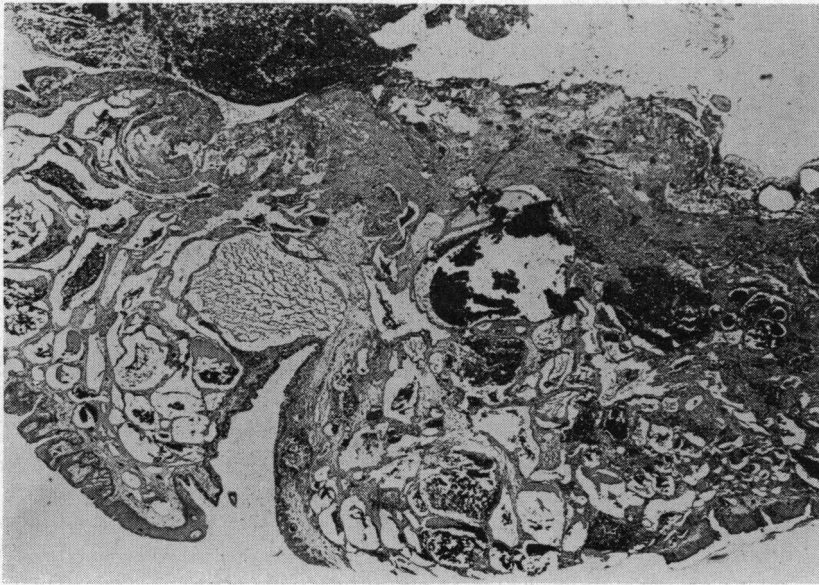


FIGURE 28. CASE 34

This specimen is from the lid portion of a lymphangioma involving orbit, lids, and conjunctiva. It is noteworthy that almost all the normal tissue has been replaced by the abnormal honeycomb growth. Several large blood channels are apparent.

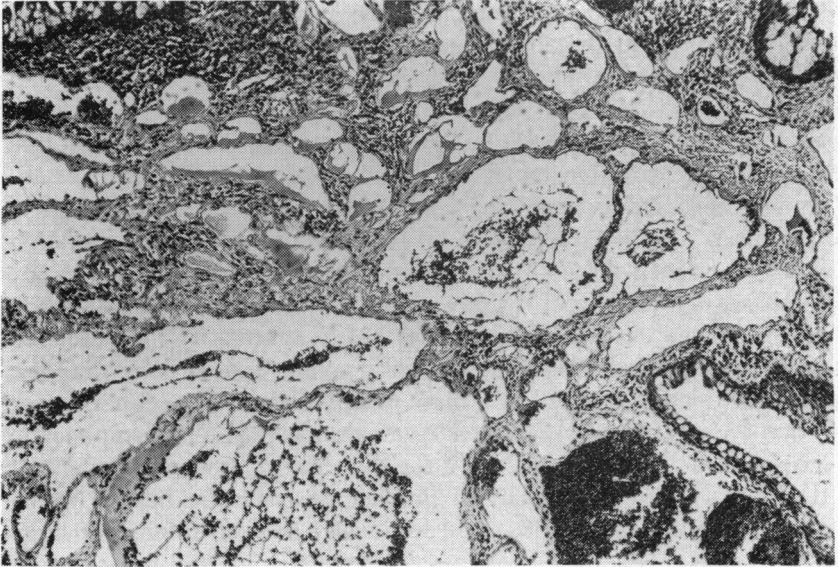


FIGURE 29. CASE 34

This is a higher power of the section in Figure 27 to show the great variation in the size of the spaces. Some of the spaces contain blood. The lymphocytic element is sparse.

vascularization following treatment with irradiation or sclerosing agents. Case 57 is an example of vascularization after sclerosing agents. It was noteworthy that those cases with heavy lymphocytic follicular formation often had the follicles projecting into the lumen of the lymphangioma spaces as shown in the section of the orbital lymphangioma, Case 37 (Figure 30).

Orbital lymphangiomas were difficult to delineate as to gross pathology because they were nearly always cystic in some portion and the collapse of the cyst during surgery altered the gross appearance. If the cystic portion of the lymphangioma did not contain blood, then the swelling was often not recognized until the cyst was inadvertently opened. At this time clear or straw-colored fluid escaped. If the lesion contained new blood or, more commonly, old degenerated blood, it was recognized as a blood cyst very early but could seldom be removed without breaking the cyst. Accordingly, most of the gross specimens of orbital lymphangioma consisted of a section of cyst wall. In the case in which exenteration of the orbit was done, the lymphangioma appeared to lie in close proximity to the short, posterior ciliary vessels. Microscopically, the lymphangioma spaces were characterized

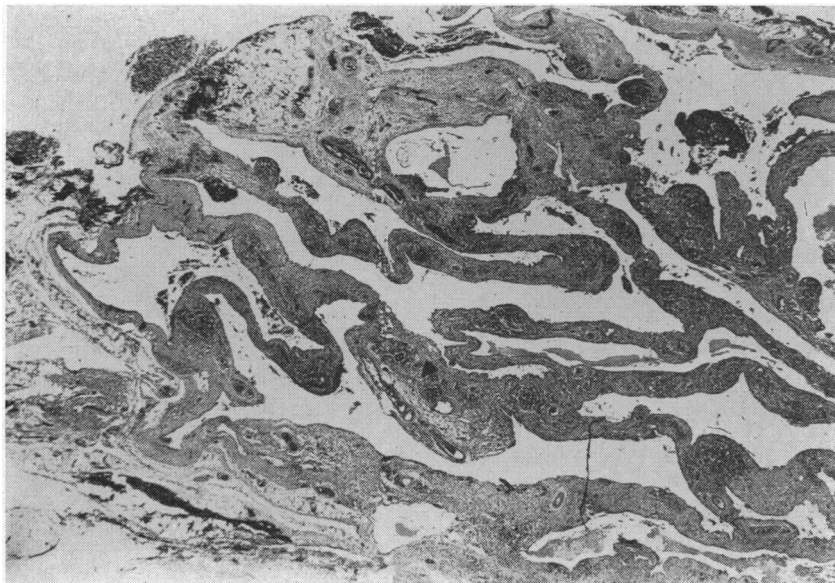


FIGURE 30. CASE 37

This orbital cystic lymphangioma demonstrates very large distensible spaces. There is a heavy lymphocytic infiltration. Some aggregations of lymphocytes project into the spaces as follicles. Many blood vessels lie in the septae, poorly insulated from the lymphangioma spaces.

by large, irregular size. Portions of the lesions showed spaces which fell in the size category of cavernous lymphangiomas, but there was usually a cystic lymphangioma portion. The amount of stroma was quite variable and the proportion of lymphangioma space to stroma was difficult to ascertain because of the collapsed nature of the space. Large blood vessels poorly supported near the endothelial lining were a common feature. Degenerated blood and blood pigment often appeared in the spaces and in the tissues comprising the wall of these lymphangiomas. The lymphocytic element was extremely variable, but heavy infiltration with follicle formation was common. The sections of Cases 39, 43, and 48 (Figures 31, 32, 33) show the foregoing features.

The pathology of lymphangiomas gives nothing which is invariably characteristic of these lesions and not found in any other condition. The picture is most often confused with hemangioma. If the spaces previously described are found to contain red blood cells, often this is taken to indicate it is a hemangioma. If the spaces are small and inconspicuous and the lymphocytic element is large, then there may

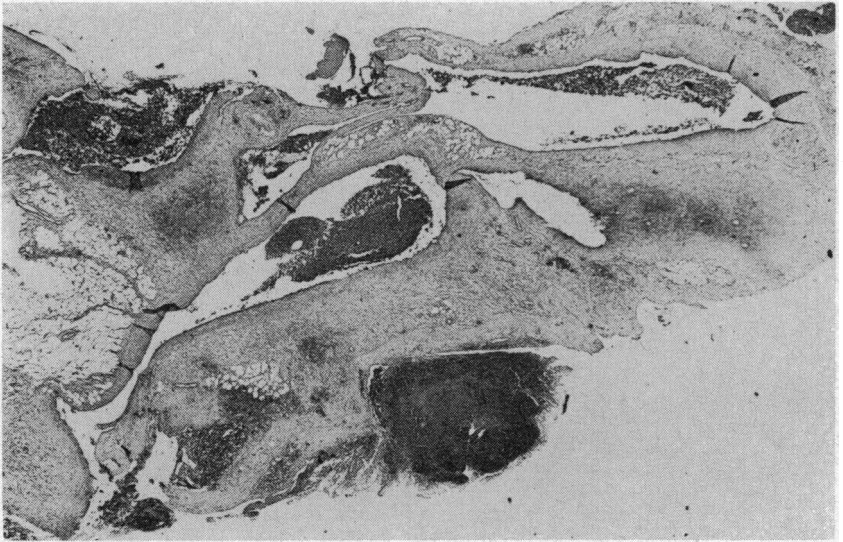


FIGURE 31. CASE 39

This was an orbital lymphangioma of long standing which was unappreciated until hemorrhage into it caused a sudden proptosis. The spaces are large and some contain blood. The stroma is heavy. The lymphocytic infiltration is slight.

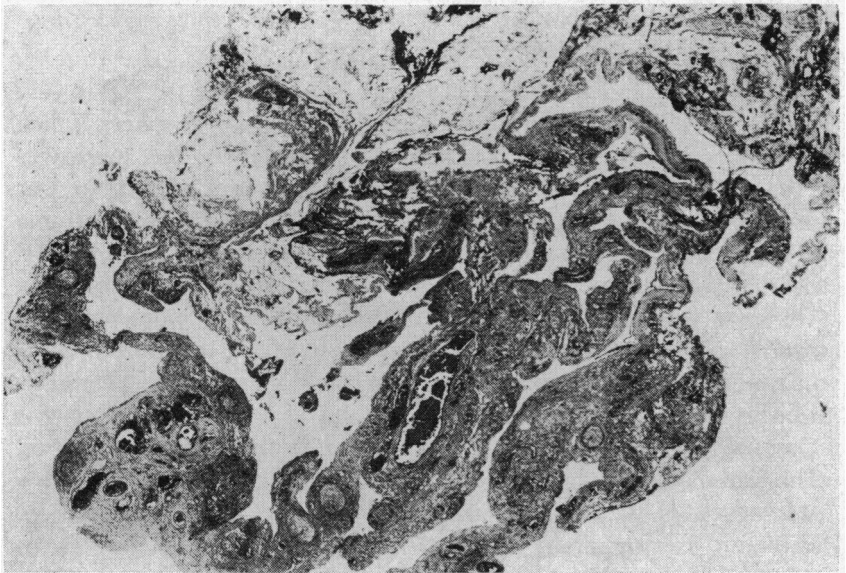


FIGURE 32. CASE 43

This was an orbital cystic lymphangioma which contained old degenerated blood. The septae are quite thick and show lymphocytes aggregated into follicles. Large blood channels in close proximity to the lymph spaces are a prominent feature.

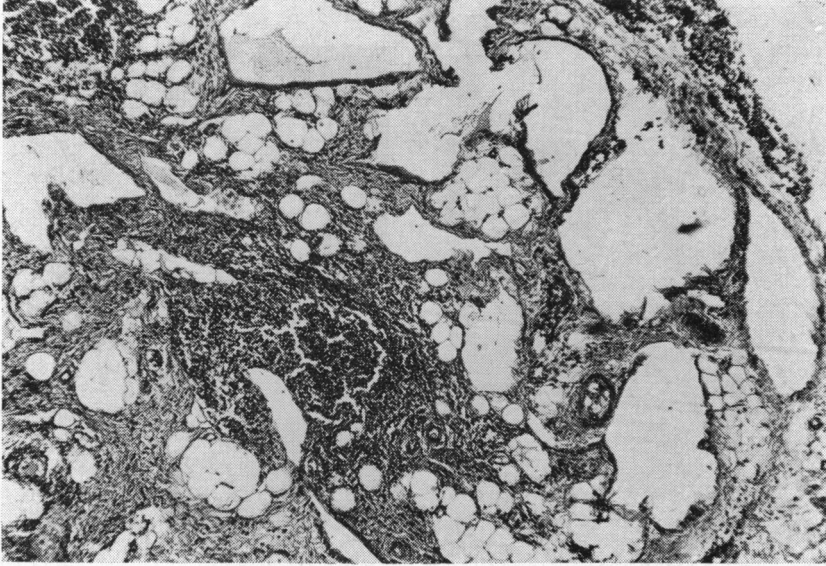


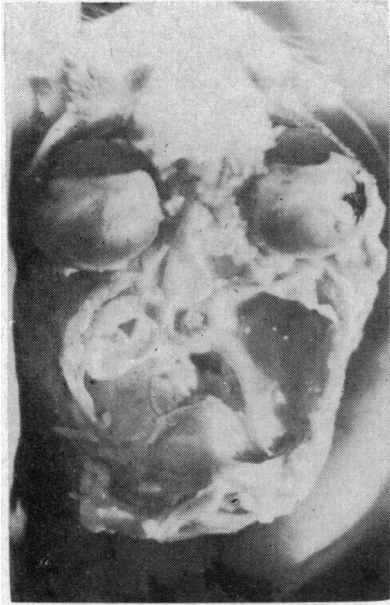
FIGURE 33. CASE 48

This case was grossly a blood cyst of the orbit which proved to be lymphangiomaticous. The specimen shows a moderate stroma containing many small spaces. Lymph follicles can be seen. The endothelial lining has been lost in many areas.

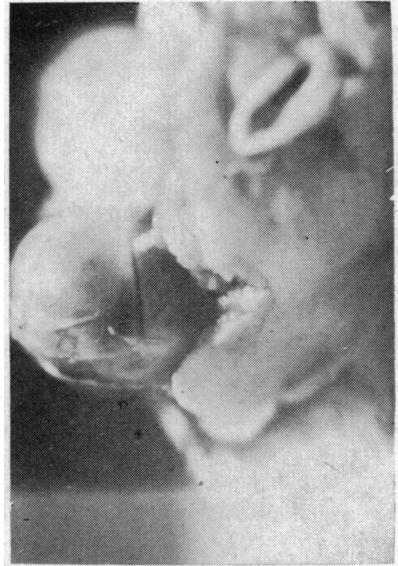
be a diagnosis of nonspecific granuloma. In those instances in which the endothelium has been lost from the lining spaces and in which old degenerated blood is present, a diagnosis of blood cyst or organized hematoma is frequently made. In general, the presence of variable-sized endothelial-lined spaces without a capsule and containing lymphocytes in the stroma is suggestive of lymphangioma. The clinical course is more characteristic of the disease than is the pathology and when taken in conjunction with the microscopic findings is usually sufficient to establish the diagnosis. Peters and Reese have described the pathology of lymphangiomas of the ocular adnexa (65, 68).

EXPERIMENTAL WORK

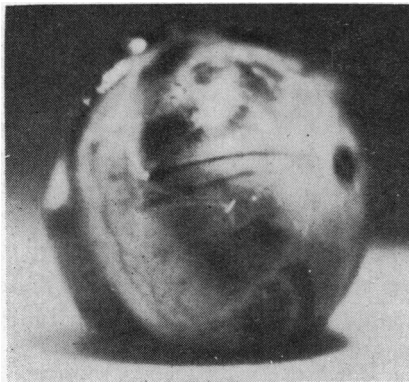
Experimental work related to lymphangiomas has taken two lines. Since it is generally thought that there are no lymphatic vessels within the orbit, and since it is equally apparent that lymphangiomas occur in the orbit, speculation arises as to whether these lymphangiomas arose from congenital rests, from backward extension of diseased or tumorous lymphatics in front of the orbit septum, or from orbital lymphatic channels. Work was undertaken to attempt to demonstrate



(a)



(b)



(c)

FIGURE 34

(a) Cat skull with the orbits unroofed, India ink is visible on the globe above on the right; (b) side view of the right globe in place, showing India ink along the side of the globe; (c) same globe enucleated. It demonstrates India ink from the epibulbar collection present in a perivascular lymphatic channel around the long posterior ciliary artery one hour after injection.

lymphatic vessels if any existed. Adult cats were used and examination of the route of disappearance of India ink after intraorbital injection was made both macroscopically and microscopically (Figure 34). Prior to injection of India ink into the orbit, 50 percent India ink aqueous solution was injected subcutaneously in the side of the cat's face and dissection was carried out to prove that the India ink was indeed picked up by the lymphatic drainage channels and carried to the regional lymph node. Subsequently, eight orbits were injected with India ink in the anesthetized animal. The orbital contents were removed at intervals of 50 min., 100 min., and 150 min. and examined under the dissecting microscope. Those globes removed after 50 min. showed a column of India ink granules in the perivascular space of the long posterior ciliary arteries. The India ink granules were moving backward in this perivascular space and had extended slightly more than one-half of the length of the long posterior ciliary vessel. It did not appear to matter whether the India ink was injected into Tenon's capsule above, below, medially, or laterally. The amount of India ink used was $\frac{2}{10}$ c.c. In those animals in which the orbital contents were removed at 100 min., it was found that the India ink column had extended the entire length of the globe and that a vascular channel containing only India ink was apparent on the optic nerve. In those animals surviving 150 min. after the injection of India ink, the granules were found in the cervical lymph nodes, but the way in which the ink got there has not yet been determined.

The second group of experiments was undertaken in monkeys to attempt to define the mechanism of the development of orbital blood cysts such as occur in cystic lymphangiomas. It was known from the present case analysis that cystic lymphangiomas were subject to episodes of sudden hemorrhage into the lesion with proptosis and ecchymosis of the lids. This proptosis then cleared more or less rapidly and in some cases did not recur. Other cases, however, without trauma, had over a period of weeks or months following the initial hemorrhagic episode a gradual increase in proptosis and at operation were found to have a cystic orbital mass containing old degenerated blood. The mechanism of the increasing proptosis in the presence of old blood without new bleeding was obscure. It seems within reason that the mechanism might be similar to that in chronic subdural hematomas. The usual explanation of the mechanism of chronic subdural hematomas is that blood enclosed within a semi-permeable membrane gains in volume by osmotic attraction as the

blood breaks down and its molecules become smaller. This increase in volume within the hematoma takes place as long as the molecules are sufficiently small to pass the membrane; then the contents of the hematoma diffuse outward and the volume is lost. Six monkey orbits were subjected to the injection of the monkey's own blood within Tenon's capsule, about $\frac{1}{2}$ c.c. being injected each time. In all instances, after the initial swelling due to the physical presence of the blood, the eye and orbit returned to normal. In the following six months none of the monkeys developed any recurrence of swelling. The next step was to place $\frac{1}{2}$ c.c. of the monkey's own blood within a cellophane envelope. This envelope was then buried within the muscle cone in eight monkey orbits. Two monkeys expired before the conclusion of the experiment. Three monkey orbits remained negative except for the initial swelling. Three of the orbits, however, had a gradual increase in prominence of the eye and orbital swelling varying from 2 mm. to 5 mm. After ten weeks the orbits were re-entered and in those monkeys with proptosis, cysts containing old, degenerated blood were found. Although this does not conclusively demonstrate that cystic lymphangiomas undergo a similar mechanism, it is none the less suggestive that the wall of the cystic lymphangioma lined with endothelium may act as a dialyzing membrane in the same way that the cellophane capsules acted in the experimental animals.

Clinical and pathologic descriptions of orbital blood cysts with suggested conformity to the above thesis may be found in reports by Awerbach (4), Augstein (1), and Golowin (29).

DIAGNOSIS

Inspection of a tumor of the ocular adnexa is often sufficient to recognize it as a lymphangioma. If there is a conjunctival portion, magnification as with the slit lamp and corneal microscope may make the diagnosis more certain.

Nearby lymphangiomas of the face, nasal cavity, paranasal sinuses, or palate may suggest the nature of a similar lid or conjunctival swelling.

The clinical behavior of the lesion, especially inexorable slow progression during the growing years, is characteristic of lymphangiomas.

Complications are often a clue to the diagnosis of lymphangioma. Repeated hemorrhage into the suspected lesion is in keeping with a lymphangioma diagnosis. Cellulitis, either with or without a con-

current upper respiratory infection, is another characteristic complication.

Refractoriness to treatment is a feature which should be counted in favor of the diagnosis of lymphangioma.

Biopsy of the lesion and examination of the tissue under the microscope may give a positive diagnosis. In those cases in which the pathology alone is not pathognomonic, combining the pathology with the clinical picture will often indicate the diagnosis.

X-ray examinations often furnish corroborative evidence. If there is a facial lymphangioma, there will be asymmetry. If the orbit is involved, most eyes will show enlargement and increased volume.

Orbital lymphangiomas without lid or conjunctival involvement probably can only be suspected until the orbit is opened at surgery. The finding of a blood cyst raises the suspicion of a cystic orbital lymphangioma, and the microscopic examination of the cyst wall will often confirm this.

The differential diagnosis of lymphangioma of the ocular adnexa concerns chiefly one other similar tumor, namely hemangioma. By inspection, lymphangiomas may appear identical to hemangiomas. In some patients mixed lymphangiomas and hemangiomas occur (Case 34). Lymphangiomas, especially after irradiation or the use of sclerosing solutions, may exhibit large racemose blood vessels (Case 57). Specimens removed for microscopic study often contain adventitious blood in the lymph spaces, making it difficult for the pathologist to choose between lymphangioma and hemangioma. The most reliable differential between these two conditions is the clinical course. The following list compares some characteristics of each disease:

	<i>Lymphangiomas</i>	<i>Hemangiomas</i>
<i>Onset</i>	Birth	Birth
<i>Course</i>	Slow progression	Rapid progression
<i>Regression</i>	Does not occur	Occurs between ages 1 and 5 (benign hypertrophic hemangioma of infancy)
<i>Complications</i>	Hemorrhage Cellulitis	Thrombosis
<i>Radiosensitivity</i>	Low	High (benign hypertrophic hemangioma of infancy)
<i>Capsule</i>	None	Present in cavernous orbital hemangiomas

The information used in the above list regarding hemangiomas is taken from Reese (68).

TREATMENT AND PROGNOSIS

Conjunctival lymphangiomas which present as clear cysts are often punctured and disappear temporarily. They usually recur, however. Since the limits of conjunctival lymphangiomas can be observed by inspection, it is usually possible to excise the entire lesion. The large number of apparent cures in the conjunctival lymphangioma cases presented herewith is indicative of this. Lymphangiomas involving the lids are less well defined and less susceptible to complete excision. Adrenal steroids have been used with improvement in elephantiasis of the lids, but it is not clear how large a proportion of the lesion was inflammatory or how great a portion was lymphangiomatous. Sclerosing solutions have brought about an improvement, but in the present group of cases no satisfactory results were obtained after sclerosing solutions and no instance of a cure could be found in the literature. Both X-ray and radium therapy likewise have effected an improvement and likewise have failed to give entirely satisfactory results. In addition, the adverse effect of radiation upon orbital bone growth in young people limits the usefulness of this approach. It has been stated by Walsh (81) that lymphangiomas are exceedingly radiosensitive. Reese (68), however, takes the view that lymphangiomas are resistant to any treatment except complete excision. Since these tumors are slow-growing, repeated partial excisions and plastic repair over a period of years may gradually get ahead of the tumor and lead to a satisfactory result as in Case 56. Orbital lymphangiomas, particularly those associated with bleeding into the spaces, form a separate category regarding treatment. If the orbital lymphangioma is susceptible to striking increase in size with inflammatory symptoms coincident with upper respiratory infection, then the anti-inflammatory measures employed against the respiratory infection will often be equally efficacious against the exophthalmos. The presence of recurrent exophthalmos together with ecchymosis indicates hemorrhage into the lymphangioma spaces which often absorbs spontaneously. Occasionally, however, the hemorrhage into the orbital lymphangioma is followed by a slow steady increase in exophthalmos lasting several months. This increase may be due to the breakdown of blood and the inhibition of fluid in a manner similar to that which occurs in chronic subdural hematomas and which was partly investigated and reported under "Experimental Work." These cases sometimes do well if the wall of the blood cyst or of the lymphangioma is widely opened allowing the lesion to collapse. Even though a complete excision is not carried out, the proptosis frequently

does not recur (Cases 38 and 57). The treatment of lymphangiomas in general is not urgent since these tumors are benign and slow-growing. Complete excision should be carried out when possible, but repeated partial excisions may be satisfactory. Drainage of blood cysts may be efficacious and anti-inflammatory measures, when indicated, are useful. Probably radiation and sclerosing solutions should not be used.

The prognosis for lymphangiomas depends upon the size of the lesion. Although the disease is present at birth, many of the smaller conjunctival lesions are not apparent until years later at which time they may be completely excised. Somewhat larger tumors may be noted early in life but may require intervention only after years of growth. Partial or piecemeal excision may be satisfactory as in Case 57. The prognosis for a cure is poor unless the lesion is small enough to be completely excised. The absence of any middle-aged or elderly patients still undergoing treatment for lymphangiomas in the present series suggests that the progression of these lesions ceases early in adulthood. Large lymphangiomas such as Case 49 and Case 53 may require exenteration. The prognosis of orbital blood cysts in lymphangiomas appears to be good even without treatment, as shown by the cases in the present series with recurrent exophthalmos and spontaneous subsidence. Removing a portion of the cyst wall seems to be curative.

GENERAL DISCUSSION

The foregoing analysis of 62 lymphangiomas shows these lesions to be benign and usually congenital. A good proportion of the smaller lesions, especially those of the bulbar conjunctiva, did not become apparent until later in life, but it is thought that some small beginning of the disease was present at birth.

The division of lymphangiomas into "capillary" and "cavernous" does not seem to have much meaning since the clinical course is the same. Designation of the cystic variety of cavernous orbital lymphangioma does indicate a clinical picture often associated with blood cysts of the orbit. Lymphangio-endotheliomas were not encountered in this series, and no comment can be made except that they are rare. The term elephantiasis is confusing when used to include lymphangiomas and probably should not be used once the diagnosis is made. Support could not be found in the cases here analyzed for a nomenclature paralleling that of hemangiomas.

The review of the literature included in this thesis is representative and is intended to be complete. Some of the citations, however, are included on presumptive grounds since the authors did not make their cases clear beyond question. There are doubtless cases of lymphangioma reported in the large body of literature on elephantiasis which are not reported here because they did not stand out with sufficient clarity. Likewise, some cases of orbital blood cyst probably deserve inclusion here but could not be added for lack of evidence in the original article.

The calculations regarding incidence of lymphangiomas are an approximation. It is probable that lymphangiomas of the ocular adnexa and face are relatively more numerous in hospital records than those of other parts of the body because their exposed position leads the patient to seek attention for cosmetic reasons.

The case reports in this thesis are all based upon microscopic examination of tissue specimens. Those cases in which complete excisions were performed and the patient cured have largely been lost to follow-up since the patients often did not return. A few of the cases presented herewith have been reported elsewhere and this is noted for each case. They are included here in order to make a complete report of all cases in the institution. As many of the cases as could be examined clinically were seen. All of the available microscopic specimens were examined. Some specimens were seen by several pathologists. If diagnosis varied, then that one which best fitted the clinical features was selected. The division of cases into three groups was on the following basis:

<i>Conjunctiva</i>	Only conjunctival involvement
<i>Lids</i>	Lids either with or without involvement of conjunctiva or face
<i>Orbit</i>	Orbit either with or without involvement of conjunctiva, lids, or face.

The proportion of orbital lymphangiomas shown in this analysis of cases is surprising. Almost half of the cases involved the orbit. It may be guessed that the rapid development of proptosis, as was the case in many of the orbital lesions, was more likely to lead to surgery than a long-standing, easily visible lid or conjunctival lesion.

The detailed analysis of these cases need not be repeated, but it is perhaps worthwhile to comment on the variations from the usual slow steady progression encountered in lymphangiomas. Perusal of hospital

records in cases of lymphangioma elsewhere than the ocular adnexa shows the most common complication to be cellulitis. Some of the cases reported here also showed cellulitis, often associated with coryza. This clinical observation coupled with the appearance of heavy lymphocytic infiltrations in microscopic preparations leads to the speculation that the cellulitis might represent a rapid heavy round cell infiltration. The second variation is spontaneous hemorrhage into a lymphangioma. This causes a sudden swelling and redness. The absorption of the hemorrhage may lead to neovascularization so that in time the lymphangioma comes to resemble a hemangioma. Cystic orbital lymphangiomas may spontaneously fill with blood, but more often do so after trauma. The association of trauma with an orbital blood cyst may lead the surgeon to drain the cyst without removing the wall for study. Possibly some cystic lymphangiomas are undiagnosed for this reason.

Although the pathology of lymphangiomas has been described, two features deserve comment. One is the presence of large poorly supported blood vessels close to the lymphangioma spaces. These vessels look prone to hemorrhage into the lymphangioma spaces. Since such a happening is common, I believe it to be a result of the precariously placed blood vessels. The second feature to be emphasized is the presence of lymphocytes often aggregated into follicles and surrounding the lymph spaces. This may explain the controversial effect of treatment discussed below.

The treatment has been discussed above and excision has been named the most effective. One of the cases improved on irradiation and later had surgery. Others did not benefit from irradiation. References are cited claiming radiosensitivity and radioresistance. It may be suggested that those cases with a heavy lymphocytic infiltration would benefit from irradiation since it is common knowledge that round cells respond well. Once the infiltration was reduced, then improvement would cease.

The literature gives little help regarding the eventual outcome of lymphangiomas of the ocular adnexa and the present series of cases also does not give the end result. The positive evidence is that some adults are unchanged or little changed since reaching adulthood (Cases 41 and 58). The negative evidence is that few middle-aged or elderly adults are represented in this series. I believe that the slow progression of lymphangiomas of the ocular adnexa ceases when growth ceases, and the lesions thereafter remain stationary.

SUMMARY

There has heretofore existed no full delineation of the character of lymphangiomas of the ocular adnexa. The present thesis is such a delineation based upon a review of the literature and an analysis of 62 cases, the largest series yet reported. The most important conclusions are listed below:

1. Lymphangiomas are congenital benign vascular tumors characterized by slow progression.
2. They differ from infantile hemangiomas in the absence of a hypertrophic phase followed by spontaneous regression.
3. Hemorrhage into lymphangiomas with or without trauma is a characteristic complication.
4. Actual infection or lymphocytosis concurrent with upper respiratory infection is common.
5. Cystic orbital lymphangiomas play a dominant role in the genesis of orbital blood cysts.
6. The mechanism of orbital blood cyst production may be identical to that of chronic subdural hematoma production.
7. Lymphangiomas are not radiosensitive.
8. The treatment of choice is excision.
9. The diffuse borders of these tumors and the lack of a capsule make complete excision difficult.
10. Small lymphangiomas respond well to treatment.
11. Large lesions are resistant to treatment.
12. The progression of lymphangiomas becomes slower as general growth slows and a stationary point is reached in early adulthood.

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