ANGIOMATOSIS RETINAE (VON HIPPEL'S DISEASE) ELEVEN YEARS AFTER IRRADIATION

BY *Frederick C. Cordes,* M.D., AND (BY INVITATION) *Ariah Schwartz,* M.D.*

In 1882 E. Fuchs (1) first described the rare disease now known as angiomatosis retinae as a traumatic arteriovenous aneurysm. Various other observers reported it subsequently under a number of different titles until von Hippel (2) established it in 1904 as an angiomatosis of the retina. In 1905 Czermak (3) described a case in association with a cyst of the cerebellum, internal hydrocephalus, and cystadenoma of both ovaries, and in 1912 Seidel (4) reported a similar case in which choked discs were observed, and later, at operation, a cerebellar cyst. It was not until 1927, however, that Lindau (5) demonstrated a close association between angiomatosis retinae and angiomatosis and cystic lesions of the central nervous system, particularly the cerebellum, and of such visceral organs as the kidneys, pancreas, ovaries, and suprarenal glands. Since then the condition has usually been referred to as the von Hippel-Lindau syndrome.

CLINICAL PICTURE

Angiomatosis retinae occurs most commonly in young adults at an average age of 25 years (6). It is bilateral in about 50 percent of cases and the tumors are multiple in some 33 percent. Hemorrhages into the retina and vitreous may also occur and are sometimes severe. There is cerebellar involvement in about 25 percent of cases and its signs usually appear some ten years after the discovery of the ocular lesions. A review of the literature indicates

* From the Department of Ophthalmology of the University of California Medical Center at San Francisco.

a definite familial incidence. The disease is usually transmitted through the female, but 60 percent of cases occur in males (7).

The fundus picture is characteristic. The retinal change first observed is a marked dilatation of one or more retinal veins. Usually a communication between a retinal artery and the affected vein can be made out; this marks the beginning of the tumor formation. From this *rete mirabile* a berry-like, reddish-colored mass develops that is sharply demarcated from the surrounding area and is supplied by an artery and a vein, both of which develop into wormlike, enlarged stems carrying dark blood. The retina surrounding the tumor is slightly elevated. Around the macula and disc, shiny white spots of exudate appear; they are at first pinhead in size but later become confluent. A globular detachment of the retina always supervenes and surrounds the enlarged tumor mass. It usually develops from the periphery and gives the impression of being semisolid. In this stage the exudate increases in amount and becomes yellowish in color. The condition progresses until there is massive detachment of the retina, marked enlargement of the vessels leading to the tumor, atrophy of the disc, and amaurosis. When the retina is completely detached it often looks as if it lay over, or as if it were permeated by coagulated milk. Iridocyclitis finally develops and is complicated by secondary glaucoma, and eventually by opacification of the lens.

PATHOLOGY

Microscopically the angioblastoma is composed of thin-walled capillaries. This would seem to provide a rationale for the use of X-ray irradiation in treating it in view of the effect that X-ray is known to have on new-formed vessels. The cells between the vessels show very few mitoses. The feeding vessels are greatly dilated and thickened, and there is a tendency for the tumor to show some cystic degeneration. Wolff (8) states that curious fat-laden cells lie between the vessels. In the usual preparation the fat has been dissolved so that the cytoplasm has a honeycombed appearance. A fibro-osteogenic membrane tends to form around the growth.

It is interesting that miliary aneurysms occurring in young

individuals, especially males, are associated with marked and progressive changes of an exudative and degenerative nature not unlike the changes in angiomatosis. Duke-Elder (9) feels that the diffuse cases actually represent a type of angiomatosis and are related to von Hippel's disease. In diabetic retinopathy a similar type of exudation has been demonstrated around some of the fine microaneurysms.

PROGNOSIS

From the foregoing it is apparent that the ocular prognosis of angiomatosis retinae is poor. This is true not only for the first eye but for the second as well, and it will be recalled that the disease is bilateral in some 50 percent of cases. There is no tendency for the tumor to metastasize. That the prognosis for life is also poor was recognized by Brandt (10) and others before Lindau showed the relationship of the eye disease to general angiomatosis. Cerebellar cysts are known to produce death in about 25 percent of cases and may do so in a considerably higher percentage since the cerebellar complications can develop after an interval of many years. Pancreatic cysts and multiple tumors in other organs have also been observed repeatedly.

THERAPY

The treatment of angiomatosis is difficult at best (Elwyn, 11) and success is possible only when the tumor can be destroyed in its early stages. No single instance of effective treatment of an advanced case has appeared in the literature. In recent years therapy has been limited to electrolysis, diathermy, irradiation with radium (radon seeds), and X-ray irradiation. It is apparent from the literature that early angiomatous growths, whether in the eye or in other structures, respond to X-ray irradiation and diathermy. So far as the retinal tumor is concerned, diathermy would seem to be as effective as X-ray but more damaging to the surrounding retina. Success with diathermy has been reported by Kaye (12), Lewis (13), Guyton and McGovern (14), and others. Moore (15) reported successful results with two patients

in whom radon seeds had been sutured to the sclera over the site of the tumor. In the first case there was no evidence of the tumor mass at the end of six months and the vessels originally supplying the growth were completely obliterated. In the second case the growths were "scarred up completely" at the end of three months.

X-ray Irradiation. Although X-ray irradiation of advanced angiomatosis has been uniformly unsuccessful, there have been a number of reports of at least temporarily successful results with early cases. Unfortunately the post-irradiation observation periods in these cases have been too short (the longest only three years) to warrant the drawing of any conclusions as to the duration of the derived benefit.

Craig, Wagener, and Kernohan (16) reported a patient to whom they gave three courses of X-ray. Three years later the large vessels leading to the mass persisted, but the angioma itself seemed to consist of scar tissue associated with atrophy and a degenerated condition of the surrounding retina. The vision of the eye at this time was 6/4.

In 1940 Cordes and Hogan (17) reported the treatment of a fairly early stage of von Hippel's disease with 1200 r. A year later the vision was still 20/30. The vessels seemed unchanged, but the exudate had markedly decreased and the tumor was much smaller and flatter. Three and a half years after irradiation (7) the vascular sacculation and tortuosity had receded but the veins remained enlarged. All signs of exudate had disappeared, the tumor mass was still elevated but smaller than before treatment, and the vision was still 20/30. Neurological examination at this time was negative except for a positive Romberg.

In 1942 Ballantyne (18) treated a case of angiomatosis retinae over a three-week period with 4,750 r divided into 14 treatments. Five months later there was marked diminution in the size of the vessels. One year after irradiation the eye showed the effects of X-ray therapy—loss of eyelashes, depigmentation of the skin of the lids, telangiectasis of the lids and bulbar conjunctiva, closure of the punctum, and general rigidity of the lids. Two years after the X-ray therapy there was an extensive intraocular hemorrhage which made enucleation necessary.

The report of Craig and Horrax (19) provided further evidence of the sensitivity of these angiomatous tumors to X-ray therapy. These observers treated a family in which three members had hemangioblastomatous growths; two showed cerebellar involvement and the third spinal cord involvement. Since operative removal of the cord lesion was impossible, the condition was treated successfully with deep roentgen therapy. In their report they referred to an earlier case treated in this way which had regained perfectly normal cord function and had retained it over a ten-year observation period.

Hirschfeld (20) reported a case of von Hippel-Lindau's disease in which there was advanced involvement of the medulla and of the retina of one eye. The patient was given a total of 1475 r directed at the medulla and recovered completely from general brain stem symptoms.

In 1941 Cordes and Dickson (7) reported a case of angiomatosis retinae in a Puerto Rican girl (K. M.); both of her eyes were involved, the right to an advanced degree, the left only moderately. Each eye was given 1800 r. Two years after irradiation the early lesion in the left eye showed improvement; the tumor had disappeared and a vision of 20/20 had been retained. During this two-year period the right eye with the more advanced process became progressively worse until there was complete detachment of the retina, gliosis, and amaurosis.

In February, 1951, M. V., a Puerto Rican girl, aged 24, was seen by one of us at the San Francisco City Hospital and was diagnosed as a case of von Hippel-Lindau's disease. She had bilateral ocular and cerebellar involvement. Investigation of her family history revealed a total of seven members, in the course of three generations on the father's side of the family, with a history of angiomatosis, either von Hippel's disease or von Hippel-Lindau's syndrome. It was learned that one of these affected relatives, an older sister, married and with four children, was the former patient, K.M., who had received X-ray therapy for angiomatosis retinae at our hands in 1941.

It is with the examination of this patient at this time, eleven years after radiation, that the present report is chiefly concerned.

OBSERVATION OF A CASE OVER AN ELEVEN-YEAR PERIOD

(December 1940 to January 1952)

RESUMÉ OF FINDINGS 1940-1942 (previously reported, 7): On December 20, 1940, K. M., a Puerto Rican girl, aged 14 years, was examined at the University of California Medical Center Eye Clinic because poor vision in her right eye was discovered during a routine school examination. (In March, 1937, she had been seen in this clinic because of "white spots" on the conjunctiva. These were diagnosed as xerosis of the conjunctiva and responded to vitamin therapy.)

Family History. The father, a Puerto Rican, had died of a brain tumor three years previously at 35 years of age. On two occasions, first in 1932 and then in 1934, he had been operated on for decompression and removal of a cerebellar neoplasm which was diagnosed as angiomatous cyst of the cerebellum. Fundus changes were compatible with increased intracranial pressure.

The paternal grandfather died "quite young" and had been blind for 9 months before death.

The mother's family history was negative.

Four sisters, aged 4, 11, 13[1], and 16 years, and two brothers, aged 9 and 12, were living and well. Examination of the fundi of all the siblings failed to reveal any abnormalities.

The general physical examination was negative.

Ophthalmic Examination: Right eye. Vision: Light perception; unimproved by lenses. Exotropia. Vitreous hazy. Disc outline almost indistinguishable because of edema and a grayish-white streak extending superiorly to a semisolid-appearing detachment of the retina. The detached retina involved the entire upper periphery and was elevated between 5 and 6 diopters. The inferior nasal vessels were dilated and their outlines became lost in a raised, cherry-red nodule about two disc diameters from the disc margin. The nodule measured three-quarters of a disc diameter. The vessels emerging from the distal side of this mass were much reduced in size and were tortuous; farther out along their course there were two other smaller reddish nodules into which both the artery and vein could be seen to enter. Superficial, soft white exudates obscured macular details. Intraocular tension was 16.5 mm. Hg (Schiötz).

Left eye. Vision: 20/20. Media clear and disc normal. A branch of the inferior temporal vein was dilated through its entire course to approximately twice its normal diameter. In the extreme periphery this vessel became lost in a reddish-yellow elevated mass one disc

[1] This was M. V. who subsequently developed von Hippel-Lindau's disease, was seen by one of us in 1951, and was the means of our rediscovery of K. M.

diameter in size. The artery leading to this mass was less dilated than the vein. There was one small, sharply defined area of exudate near the inferior nasal vein in the mid-periphery of the retina. The intraocular tension was 16.0 mm. Hg (Schiötz).

Diagnosis. Well-advanced angiomatosis retinae of the right eye with an early lesion of the same nature in the left.

Treatment. A course of X-ray irradiation was given between February 18 and March 5, 1941. Each eye received 1800 r.

In the published description of this case abstracted above, the findings on December 15, 1942, 22 months after radiation, were the last data reported. At this time the right eye showed a more extensive detachment of the retina and an increase in the amount of gliosis. The intraocular tension was 30 mm. Hg (Schiötz). The lesion in the left eye was smaller, flatter, and whiter than it was before irradiation, as though scarring had taken place.

FINDINGS, 1943 (not previously reported): On July 15, 1943, the patient was examined again. The right eye appeared not to have changed since the previous examination, but the lesion in the left eye seemed a little larger and more elevated. From August 13 to August 25, 1943, two and a half years after the original roentgen therapy had been applied, a course of X-ray totaling 2000 r was given to the left eye.

On October 11, 1943, there was no apparent change except that the tension of the right eye had risen to 40 mm. Hg (Schiötz).

The patient failed to keep her next appointment and investigation revealed that she had moved and left no forwarding address. She was not seen by us until March, 1951, when she was examined in connection with her sister's illness (see above).

FINDINGS on January 11, 1952, 11 Years after First X-ray Irradiation:

Right eye. Amaurotic. Intraocular tension: 50 mm. Hg. (Schiötz). Exotropia. Deep anterior chamber. No congestion. Mature cataract.

Left eye. Vision: 20/20. Intraocular tension: 20 mm. Hg (Schiötz). External and slit-lamp examinations negative; no sign of lens opacification. Media clear; disc and macula normal. Neither the vessels nor the angiomatous mass appeared to have changed since the examination of October, 1943. The small patch of exudate previously observed inferonasal to the disc had disappeared.

One disc diameter superotemporal to the old mass was a new lesion: a small, round, grayish-pink mass, one-eighth disc diameter in size and slightly elevated. This mass was fed by a branch of the inferior temporal artery and drained by a branch of the inferior temporal vein. These vessels were normal in size and configuration. There was a small patch of exudate between the inferior vessels but the remainder of the retina was negative.

The general physical examination at this time was negative.

The patient will be kept under observation and the advisability of further radiation for the new lesion will be weighed.

SUMMARY AND CONCLUSIONS

1. Angiomatosis retinae (von Hippel's disease) is a rare, progressive disease characterized by a vascular tumor which in its late stage is marked by gross enlargement of the vessels leading to it, massive detachment of the retina, atrophy of the disc, and amaurosis; it is frequently associated with angiomatosis and cystic lesions of the central nervous system and viscera (von Hippel-Lindau syndrome).

2. Both eyes of a case with an early lesion in the left eye and a well-advanced lesion in the right eye were treated with 1800 r; two years later the left eye was exposed to an additional 2000 r. Eleven years after the first course of X-ray irradiation the well-advanced lesion of the amaurotic right eye remained unchanged; the lesion in the left eye showed atrophy and scarring but no evidence of progression or of lens opacification, and vision was still 20/20.

3. Although the tumor in the left eye had responded to radiation therapy, there was no change in the enlarged vessels leading to it.

4. A small new angiomatous growth, not present when the patient was seen nine years earlier, had developed in the left eye.

5. The results of this 11-year follow-up, which was 8 years longer than any other post-irradiation observation period on record, confirmed previously recorded experience that treatment of angiomatosis retinae with X-ray irradiation is of no avail in advanced cases but may be successful in early cases.

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DISCUSSION

DR. ALGERNON B. REESE. Dr. Cordes has given us a most unusual and well-documented case of angiomatosis retinae. Seven members of a family in three generations had the disease. The patient had four children, and I am sure that Dr. Cordes has examined or will examine their fundi in view of the fact that the disease is transmitted through the female. He states one sister was examined in 1940, and the fundi were thought to be normal. Later this patient showed signs of the disease. I should like to ask Dr. Cordes if he does not feel that at this previous examination at least some degree of a matrix of the lesion was present in view of the fact that it is generally felt the condition is congenital.

Should these cases of angiomatosis retinae be treated? I do not believe there is any doubt that they should be because, although the tumor is not a metastogenic one, it is certainly locally destructive to the eye. The question then arises as to the mode of treatment. There are two types available, one diathermy, and the other, some form of irradiation. Diathermy has the advantage in that it can be applied locally to the site of the lesion to the exclusion of the tissue elsewhere. It is true

a needle might injure one of the large efferent or afferent vessels and produce hemorrhage in the vitreous. I know of no instance in which this has occurred. Irradiation in some form, preferably X-rays, has the disadvantage in that the rays certainly affect to some degree the entire posterior sector of the eye to say the least, and they are given in the hope that they will have a selective action on the tumor. Some damage is done, in most instances minimal, to the normal structures of the eye. One of the complications I should like to mention is a late vitreous hemorrhage that occurs maybe a year and a half or two years following irradiation. Ballantyne's case, quoted by Dr. Cordes, had such a hemorrhage a year and a half after treatment. Presumably the X-rays cause changes which lead to occlusion of a retinal vein from which the vitreous hemorrhage occurs.

I would suggest, therefore, the following rule in the treatment of these lesions—the diffuse extensive, multiple ones be treated by X-rays and the localized, single lesions be treated by diathermy. I have treated a case by diathermy and there has been no progression now for seven years.

I have been rather skeptical as to the efficacy of irradiation in these cases, but Dr. Cordes has convinced me that it can be an effective method of treatment. I personally favor diathermy when it is possible to use it for the reasons given, and I think Dr. Cordes should use diathermy to the small area of recurrence which he has noted in the case he reports.

DR. DERRICK VAIL. I would like to compliment Dr. Cordes on a most wonderful presentation in his usual masterly fashion. I should also like to repeat, perhaps in a little different words, the remarks of my predecessor, Dr. Reese, in discussing this subject. In 1946 I had the privilege of seeing a case of angiomatosis retinae in a woman of about 30 years of age.

This picture shows the lesion in the lower temporal area surrounded by some edema of the retina, and a peculiar flaky lipoid (if it is lipoid) deposit on the basal side of the tumor, between the tumor and the foveal zone. A diathermy coagulation was performed, and the second slide shows the picture of the tumor with the lipoid deposit. The third slide shows the peculiar fluffy exudate which has been described in all these cases. As I stated, diathermy coagulation was performed in the lesion, and the result was excellent.

The next slide depicts the drawing of the fundus 6 months after the lesion was treated, showing the area well walled-off and destroyed. About a year later the lipoid substance had disappeared. The vision improved from 20/70 at the onset to 20/20 at the end of the year, and recent studies of the patient show an entirely healed lesion. I should

like therefore to emphasize what Dr. Reese has said: when the area is sharply defined it is not difficult to localize the lesion and to treat it properly with diathermy. I would also suggest doing a few punctures into the tumor itself if surface diathermy be applied in the area. This may possibly prevent the hemorrhages that have been mentioned. I will leave it to Dr. Guyton to describe his case where hemorrhage resulted in the loss of the eye.

DR. RALPH I. LLOYD. I have the privilege of reporting a case under observation for 20 years.

The first slide shows the eye of a girl of 16 who came to the Brooklyn Eye and Ear Hospital in 1931. Dr. Evans and I have kept her under observation up to 1951. This left eye of course is out of commission. I particularly call to your attention the quiescent lesion above the macula which remained as long as we could observe the interior of the eye. The main lesion was so far forward you could not see it.

The other eye had a vison of 20/20. The lesion is quiescent at this time and we observed it for a while with the idea it would behave as did the one in the other eye.

A picture taken 8 months later shows marked dilatation of both artery and vein connected with the angioma and evident involvement of the macular area as shown by little spots of exudate.

The next picture was taken a few months later. This time we consulted a reliable radiotherapist. He did not advise therapy because of proximity to the macula. If we had known what was to happen, we might have recommended diathermy puncture despite the risk.

The next picture shows the eye about 2 years later with an angry lesion present forward and the central vision absent.

The subsequent history of the case is interesting.

In 1936, the young woman became pregnant; to avoid strain on the vascular system a Caesarian section was done. About as close after that as was possible another Caesarian was necessary, which she survived. In 1941 she developed symptoms of dizziness and headache, and Dr. E. Jefferson Browder removed a cyst from the left cerebellar lobe. The tissue removed gave no clue of the type. It was a large cyst with no blood vessels in the wall.

The patient was last seen in 1951. Both eyes were blind, the tension elevated, the corneas degenerated, and she was confined to a tuberculosis institution.

The last slide shows the differential diagnosis between arteriovenous communication of the congenital type and angiomatosis. The vision in this case is 20/20 and both arteries and veins involved retain their normal features despite enlargements resulting from short-circuiting the circulation. In angiomatosis artery and vein lose their distinguish-

ing features and you can only tell which is artery and which is vein by tracing them back to the disc.

DR. PHILIP M. LEWIS. Dr. Cordes is to be congratulated on the excellence of his result in this case. It certainly shows that the disease can be successfully treated by means of irradiation. However, in most cases of angiomatosis retinae the treatment of choice is electrocoagulation. Two cases that were successfully treated by electrocoagulation have been under my observation for ten and one half and eight years respectively. In the first case permanent damage to the macular function had occurred prior to surgery. However, the eye is not completely blind, maintaining over the years a vision of about 4/200. Even the preservation of a globe in these cases is important to the patient. There was much scarring and atrophy, as will be shown in the slides.

The first slide shows the fundus with the very large vessels, taken before the electrocoagulation. The second slide shows the macular region, with sheets of degeneration which had already taken place and which doomed the vision of the eye permanently. The third picture shows deposits on one of the large vessels.

The first picture taken after surgery shows the scarring, atrophy, and pigmentation.

These pictures were taken over the years. The later ones show that the large vessels have entirely disappeared, and that there is a great deal of permanent scarring.

The second case was seen early in the wards of the John Gaston Hospital. The patient came in because of severe headaches, and we were afraid she might have cerebellar involvement or some tumor of the brain. This was not the case.

The slide shows large vessels coursing on down to the angioma, which may be seen below in the six o'clock meridian.

The first picture taken after surgery and subsequent pictures reveal that remnants of the vessels were still visible, and there was scarring at the site of the angioma.

These cases were both reported before this Society in 1947. It happened that both growths were in the periphery where they could be easily attacked. This last case has maintained a vision of 20/20 and Jaeger I, through the years, and I feel will continue to do so. Her only defect is a notching in the superior field of which she is unaware.

The term "amaurosis" which Dr. Cordes used in speaking of the blind eye of his patient seems a little confusing to me. While it is not incorrect, I think it should be reserved for those cases which are blind without demonstrable fundus pathology.

In closing I should like to stress what Dr. Cordes has already em-

phasized, namely that these cases should be treated early in order that vision may be preserved.

DR. F. C. CORDES. In the presentation no attempt was made to evaluate the various methods that have been employed in the therapy of angiomatosis retinae. Doctor Reese suggested that X-ray might be indicated in those cases where there are multiple lesions and where these lesions are located in an area close to fixation. Here it would be most essential to keep the destructive action on the retina down to a minimum and perhaps this would be the area where X-ray therapy would be justified, even though there be a risk of developing a radiation cataract. On the other hand, if the lesion is in the periphery it would seem that diathermy is probably the method of choice in destroying the tumor. In the case described here, which has already received 3800 r, and where additional X-ray therapy should be avoided, diathermy would be the treatment of choice.

I want to thank the discussers for the interest that they have added to this case presentation.