

THE BRITISH JOURNAL

OF

OPHTHALMOLOGY

JULY, 1939

COMMUNICATIONS

CHOROIDAL ANGIO-SCLEROSIS WITH SPECIAL REFERENCE TO ITS HEREDITARY CHARACTER

BY

ARNOLD SORSBY

LONDON

(1) Review of Literature

UNDER the name central senile areolar choroidal atrophy, Nettleship described in 1884 a bilateral affection leading to grave impairment of vision. In a relatively sharply circumscribed area extending from the disc to well beyond the macula, varying stages of atrophy of the choroidal vessels could be seen; in the left eye the lesion was almost perfectly circular, in the right rather larger and not quite so regular in outline. The patient was aged 60 years and her sight had been failing for 12 years; her father had been blind six years before he died, but no details were known.

In his report, Nettleship drew attention to a description by Jaeger in 1855. Jaeger's case (described as disappearance of the pigment epithelium of the choroid at the macula lutea) appears to have been unilateral, and as the choroidal vessels are not shown as white lines, Nettleship's case is more definite and more instructive. An almost parallel case was described by Retze in 1902, whose patient, a man aged 66 years, had failing sight for two years and showed the same circumscribed circular area of sclerosis

extending temporally from the disc to well beyond the macula and engulfing it. In both these cases the exposed choroidal vessels revealed almost every range from the normal red colour to conversion into white streaks. A rather similar appearance was reported by Hugh Thompson (1905) in a woman aged 63 years. More recently François (1936) described two such cases.

A case of some interest is described by Knapp under the name of *sclerosis choroidea circinata*.—In a girl aged 11 years an oval area surrounding the macula in each eye assumed the appearance of a *caput medusae* owing to definite sclerosis of the choroidal vessels seen at the periphery of the oval and most marked towards the disc; the macular region itself appeared fairly normal, but the foveal reflex was absent. The family history was negative except for the fact that the grandparents were cousins. The patient herself was healthy. There was night blindness and a scotoma corresponding to the area affected.

No other illustrated descriptions of cases of the kind described by Nettleship and Knapp appear to have been published, but case reports of a number of rather similar conditions are available.

Haab's Atlas contains an illustration of choroidal sclerosis surrounding the disc and extending towards the equatorial region; two somewhat similar cases are illustrated by Di Marzio. Patchy sclerosis near the disc on a background suggestive of mild generalised choroidal sclerosis is pictured by Oeller in healthy patients aged 57 and 71 years. Under the name senile choroiditis Frost's atlas illustrates a case in a man aged 83 years. More extensive lesions are reported by Levinsohn and by Bednarski in whose cases there was widespread, rather patchy, atrophy of retina and choroid in addition to fairly generalised choroidal sclerosis. Levinsohn's patient was a man aged 60 years, of non-consanguineous origin, and healthy apart from his visual defect which had developed seven years before and was associated with night blindness; the family history was clear. Bednarski's case referred to a man aged 23 years, one of a family of 10 of whom 6 died of unknown causes in infancy. The patient developed night blindness at age of 8 years and vision had been failing since. To Bednarski the vascular changes and the rather irregular pigmentary degeneration suggested a mixture of two clinical conditions: choroidal sclerosis and gyrate atrophy of retina and choroid. A case reported by Holloway (1914) appears to have been rather similar to Levinsohn's. More recently Pillat reported two cases of this kind under the name of tapeto-retinal degeneration of the central fundus region. His patients were males aged 42 and 51 years with no history of consanguinity. White dots were present in the periphery of the fundi and were regarded as evidence of retinitis punctata albescens, the central lesion itself

being regarded as central retinitis pigmentosa. Some peripheral loss of field was present in one case.

Sclerosis involving practically the whole of the choroid has been reported by a number of observers. Three plates in Jaeger are highly suggestive, but the first definite case comes from Stanford Morton, whose patient, a man aged 55 years, had full central vision, but a field reduced to 10° . Except for the macular area, the retinal pigment layer and the chorio-capillaris were absent all over the fundus; the sclerosis was most marked towards the disc, the peripheral choroidal vessels appearing normal; pigmentary disturbances were of the slightest. Increasing night blindness and failing vision had been present for about five years. Two plates of rather similar conditions appear in Frost's atlas. A later report came from Bishop Harman, whose patient, a woman aged 57 years, had upward coloboma of the iris in each eye; sight had been failing for 24 years. A case of Thompson (1901) may perhaps also belong to this group. Weiss, reported in abstract, appears to have demonstrated a child with congenital unilateral complete sclerosis of the choroidal vessels.

Aetiological Considerations :

1. *Age.*—Most patients with choroidal sclerosis, no matter of what type, were elderly. But in some cases the history was of long duration, as in Harman's case; Bednarski's patient was aged 23 years and developed night blindness at 8 years; Knapp's patient was aged 11 years.

2. *Syphilis.*—In a case of rather patchy distribution illustrated Oeller (b), and in one of Di Marzio's patients, syphilis appears to have been present. A syphilitic aetiology is suggested in another case of Oeller (a) (1899) and was assumed by Guglianetti for a case of generalised choroidal sclerosis.

3. *Tuberculosis.*—Pulmonary tuberculosis was assumed to be the cause in one of Di Marzio's cases.

4. *Cardio-vascular disease.*—Only in one case (Harman 2nd case) was there a definite heart lesion. Most reports speak of the patient's good health. Morton reported of his patient that seven years later the general condition had deteriorated and that there was albuminuria.

5. *Familial Incidence.*—Most case reports give a negative family history; a familial factor is suggested, but not conclusively, in four case histories.

(i) Holloway and Fewell (1919). In a high myope aged 53 years, whose vision had been poor since childhood, but

who could read till about the age of 27 years, there was generalised choroidal sclerosis, most marked centrally. There was no consanguinity; four brothers living all had poor sight; of six children four survived and had good vision.

(ii) Cuperus (1903). This report is rather more conclusive. In a man aged 70 years, there was choroidal sclerosis radiating from the disc and extending towards the equator. Vision had been failing since the age of 30 years. The parents could see well, but a grandmother died blind. One brother died blind, and another still alive was blind. The patient stated that visual defect in the affected members of the family always developed at about 30 years. According to the report by another ophthalmologist the blind brother alive showed the same fundus appearances as those described by Cuperus for his patient.

(iii) Guglianetti (1908). A man aged 32 years showed widespread choroidal sclerosis with some discrete pigmentary disturbances. The periphery of the fundus was normal. There was no consanguinity and no history of blindness in the family. The patient was subject to Raynaud's disease. According to the report by another oculist, the fundus appearances were the same in a sister, aged 31 years; she was not subject to Raynaud's disease.

(iv) Wilmer (1934). A man aged 35 years with a record of "retinitis pigmentosa" for four generations on the maternal side, the mother herself having no eye defect, showed extensive choroidal sclerosis. Pigmentary disturbance was not a marked feature. Judging by the family tree given by Wilmer, the patient had no brothers or sisters and the four ascendants who were supposed to have been blind from retinitis pigmentosa were all males.

In addition to these four groups, two further observations deserve notice:—

(i) Gilbert (1930), in the section on the choroid in the *Kurzes Handbuch der Ophthalmologie* illustrates a case of choroidal sclerosis and states in the text that "familial aspects, hereditary inferiority of the vascular system, are occasionally unmistakable." No further details are given.

(ii) Kapuscinski (1934), reports a family with three affected sibs but normal non-consanguineous parents, showing symptoms suggestive of Friedreich's ataxia, mental deficiency, hypogonitalism together with myopia and choroidal sclerosis

(of the peripapillary type). This group bears some resemblance to the Laurence-Biedl Syndrome and is discussed elsewhere (Sorsby, Avery and Cockayne, 1939).*

6. *Relationship to retinitis pigmentosa*.—In none of the reported cases of choroidal sclerosis was pigmentary disturbance a marked feature, though some clusters of pigment were present in practically all cases. Nor was the appearance of the discs, ranging from normality to atrophy, suggestive of retinitis pigmentosa, though Holloway reports some yellowish pallor of the discs in his case. The retinal vessels were normal in most cases, or showed relatively little narrowing except for marked constriction noted in the case of Holloway and Fewell. Reduced light sense or actual night blindness is noted by most observers. The fields in cases of generalised sclerosis showed reduction to the fixation spot (Morton, Frost) or complete extinction (Harman), but in the cases of localised sclerosis, the field defect corresponded to the affected area, except for some peripheral limitation in the cases of Levinsohn and Pillat.

The possible relationship of choroidal sclerosis to retinitis pigmentosa is further brought out by two families described by Pöllot (1911) and by Di Marzio (1938). Under the name of hereditary atypical retinitis pigmentosa, Pöllot described a boy aged 17 years, showing fairly generalised choroidal sclerosis; his mother had night blindness, and peripheral pigment changes similar to those seen in retinitis pigmentosa. A cousin also showed choroidal sclerosis, and his mother had typical retinitis pigmentosa; a brother was affected with night blindness. Some resemblance to this family with retinitis pigmentosa showing heavy choroidal involvement, is seen in the report by Di Marzio. The patient, a man aged 45 years, had been blind from infancy;

* (1) Gilbert's observation is now extended by a brief account of choroidal sclerosis in two brothers aged 44 and 45 years. (*Ber. deut. Ophthal. Gesellsch.*, Vol. LII, p. 447, 1938.)

(2) Earlier references to familial choroidal sclerosis appear to be contained in the following observations:—

(a) Hutchinson, J. (*Ophthal. Hosp. Repts.*, Vol. VIII, p. 231, 1875.) In his classical account of what is sometimes known as Tay's choroiditis, Hutchinson described amongst other cases a familial group of central lesions in the fundus. These cases have been regarded as the first observations of Doyne's choroiditis (antedating Doyne) and alternatively as cases of macular dystrophy. It is difficult to make a convincing retrospective diagnosis on the description (devoid of illustrations), but it is quite possible that the patients were examples of familial central choroidal sclerosis.

(b) Magers in a Thesis, Jena, 1899 (quoted by Leber in *Handb. der ges. Augenheilk.*, Leipzig, 2te Auf., Bd. 7 ii, S. 1208), probably saw a familial group.

(c) Haas (*Nederl. Tydschr. v. Geneesk.*, Vol. LXXV, p. 4720, 1931) reports a familial group of macular dystrophy, but his cases undoubtedly represent central choroidal sclerosis. (A man aged 80 years and three of his sons aged 53, 51 and 48 years were affected; two younger daughters aged 46 and 36 years and a son aged 35 years were normal.)

an illustration of the fundus does not show the sclerosis mentioned in the text; a brother and a son are said to be blind. Di Marzio considers this case as one of congenital choroidal sclerosis and an anomalous form of retinitis pigmentosa.

Summary

1. It would appear that three clinical forms of choroidal sclerosis have been described.

(a) "Central senile areolar choroiditis" (Nettleship, Retze, Thompson). In this type a more or less regular oval area extends from the disc temporally and engulfs the macula.

(b) Peripapillary type (Haab, Harman, Cuperus, Guglianetti, Di Marzio, Pillat). Here the choroidal sclerosis radiates from the disc to a variable but considerable extent peripherally in all directions.

(c) Generalised sclerosis (Morton, Frost, Harman). In Morton's and Frost's cases the macula was spared allowing good central vision, with fields of a tubular type. In Harman's case the macula was engulfed.

2. Evidence of syphilis was present in four cases (Oeller, Frost, Guglianetti, Di Marzio); and of tuberculosis in one (Di Marzio). Otherwise the case reports are negative, except that remote consanguinity is noted by Knapp and a possible familial factor is suggested by Holloway and Fewell, by Cuperus, Guglianetti and by Wilmer. In practically all cases, those of Pöllot and of Di Marzio excepted, the diagnosis of "atypical retinitis pigmentosa" could be ruled out.

(In this review the limited peripapillary choroidal sclerosis sometimes seen in myopia and the patchy secondary sclerosis following inflammatory lesions in the choroid have not been considered.)

(2) Case Reports of Familial Choroidal Sclerosis*

(i) "CENTRAL SENILE AREOLAR CHOROIDAL ATROPHY." George R., aged 59 years. Vision: 3/60 each eye. Peripheral fields full. No night blindness. Up to about the age of 20 years was not aware of any eye trouble. Sight has been getting worse progressively till about 10 years ago. A complete medical examination is entirely negative. (W.R.: negative. Electrocardiograph: normal.) Fundi show sharply defined "areolar atrophy" with

* Most of the patients described here have been demonstrated at different times before the Ophthalmological Section of the Royal Society of Medicine. (Meetings of January 1935, December 1936, March 1938, and November 1938).

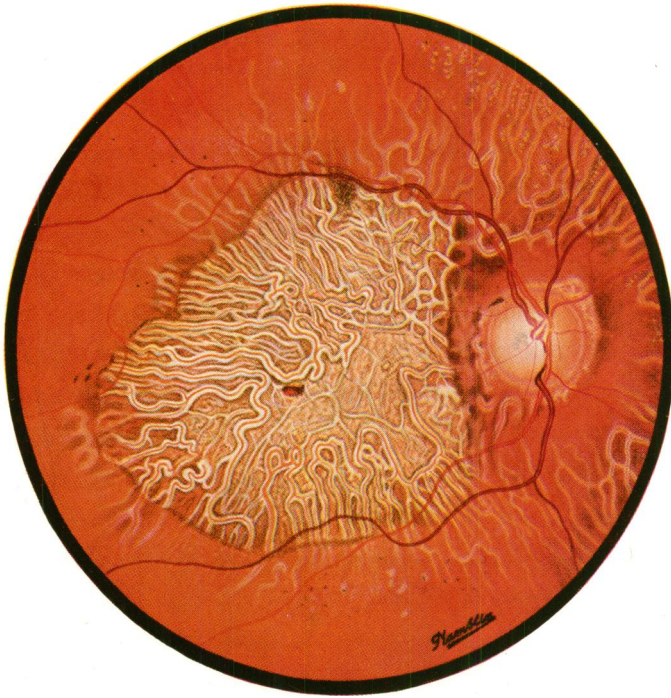


FIG. 1.

“Central senile areolar choroidal atrophy.” Note the extensive choroidal sclerosis and the sharp demarcation of the lesion.

Fig. 2 shows the appearances in the other eye.

Figs. 3 and 4 illustrate the changes seen in the patient's brother.

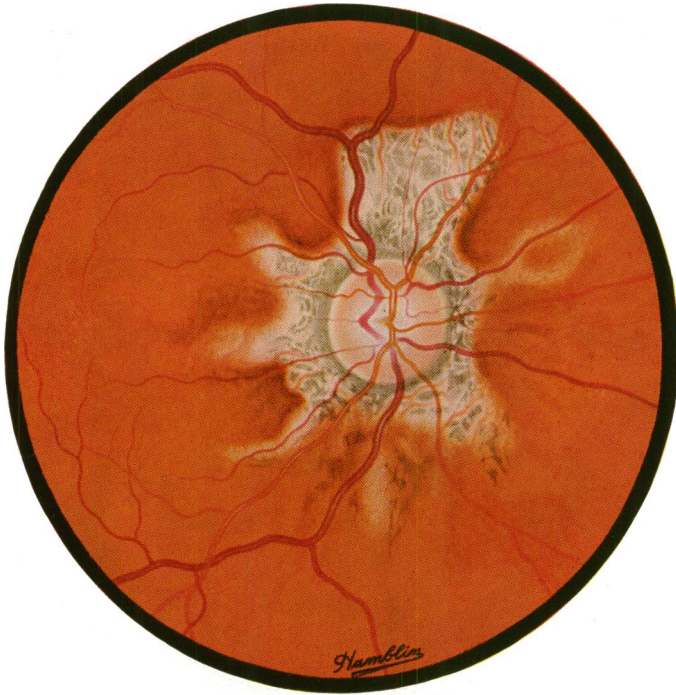


FIG. 5.

Peripapillary choroidal sclerosis. Note the limited distribution of the choroidal sclerosis around the disc. Central vision was unaffected in this eye.

Figs. 6, 7 and 8 illustrate the rather more extensive peripapillary choroidal sclerosis in the other eye, the presence of choroidal haemorrhage, its absorption and the progressive disappearance of sclerosed choroidal vessels.

Figs. 9 and 10 illustrate the more extensive lesion seen in the sister in whom the central areas have become engulfed.

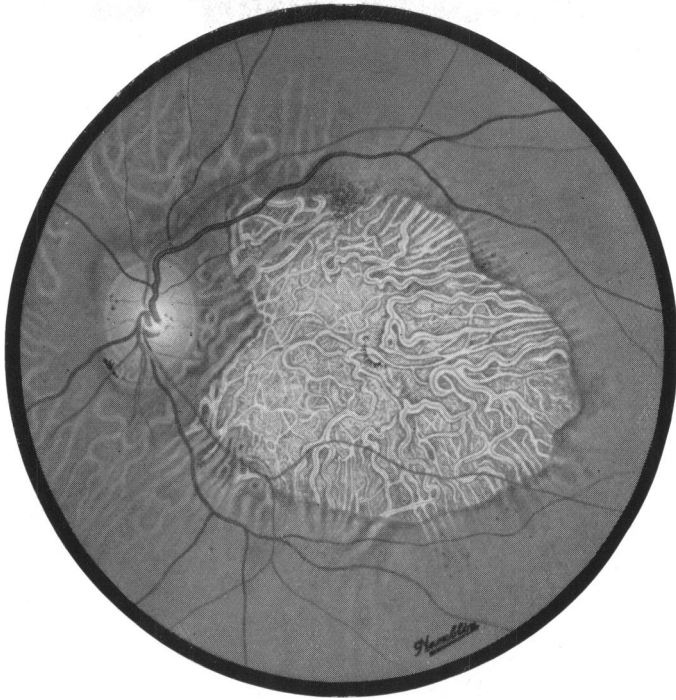


FIG. 2. For legend see Fig. 1.

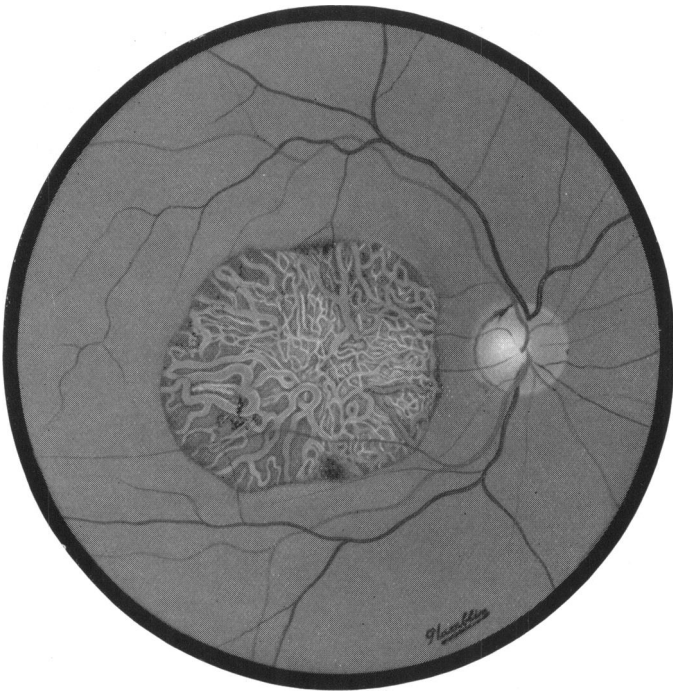


FIG. 3. For legend see Fig. 1.

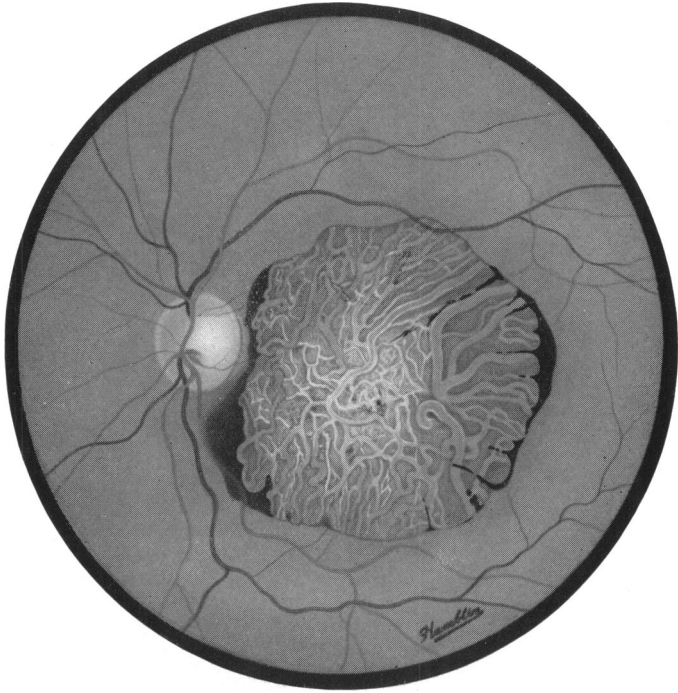


FIG. 4. For legend see Fig. 1.

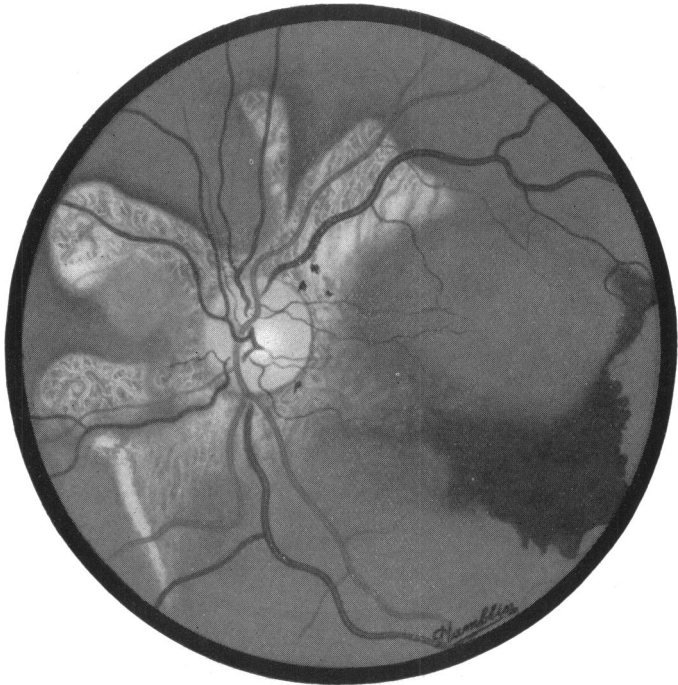


FIG. 6. For legend see Fig. 5.



FIG. 7. For legend see Fig. 5.

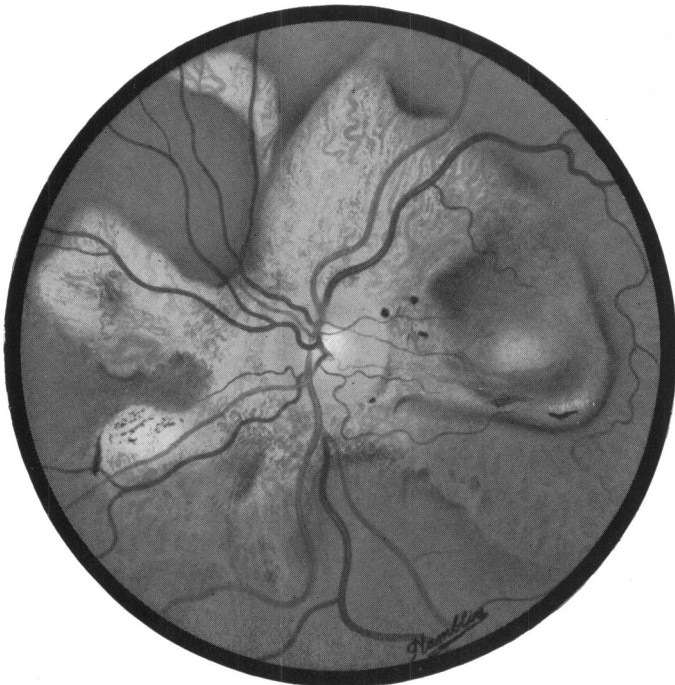


FIG. 8. For legend see Fig. 5.



FIG. 9. For legend see Fig. 5.



FIG. 10. For legend see Fig. 5.



FIG. 12. For legend see Fig. 11.



FIG. 13. For legend see Fig. 11.



FIG. 11.

Central and peripapillary choroidal sclerosis. The lesion here bears some similarity to those illustrated in Figs. 9 and 10, at any rate as far as extent is concerned. Yet the lesion in this eye, in the fellow eye shown in Fig. 12 and in the sister's eye (Figs. 13 and 14) does not appear to have originated as a peripapillary choroidal sclerosis extending and engulfing the central area. In these patients the lesion seems to have started in the central area as is shown by Fig. 15 which illustrates the changes seen in 1926 in the same eye as shown in Fig. 11.

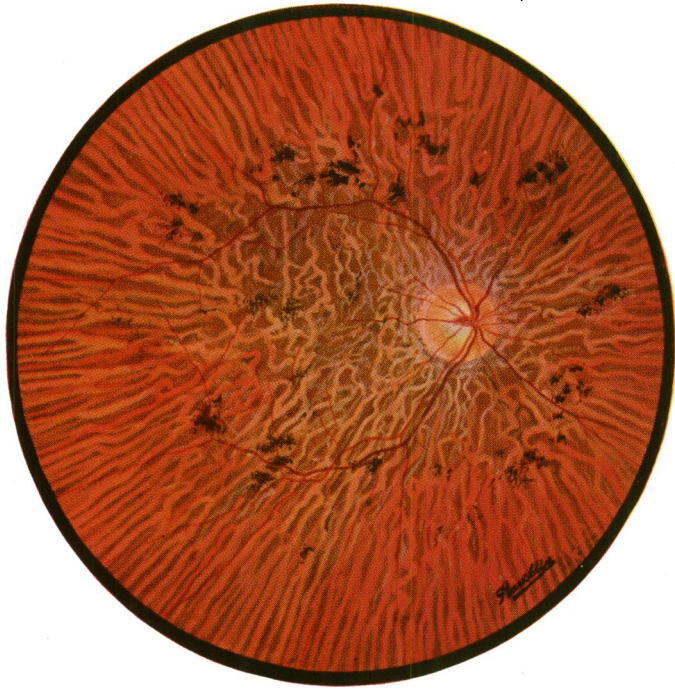


FIG. 16.

Generalised choroidal sclerosis. Note the generalised choroidal sclerosis, most marked around the disc. The pigment disturbances bear some similarity to those seen in retinitis pigmentosa, but the disproportion between the vascular and the pigment changes is striking. Fig. 17 shows the appearances in the other eye and Figs. 18 and 19 in those of the patient's sister.



FIG. 14 For legend see Fig. 11.

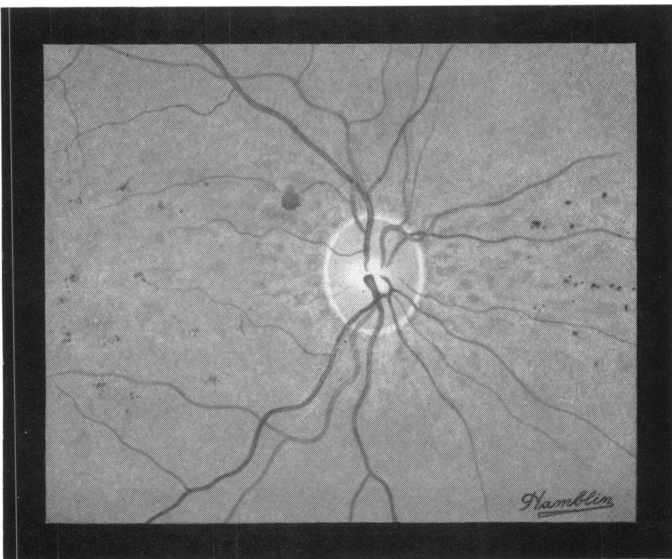


FIG. 15 For legend see Fig. 11.

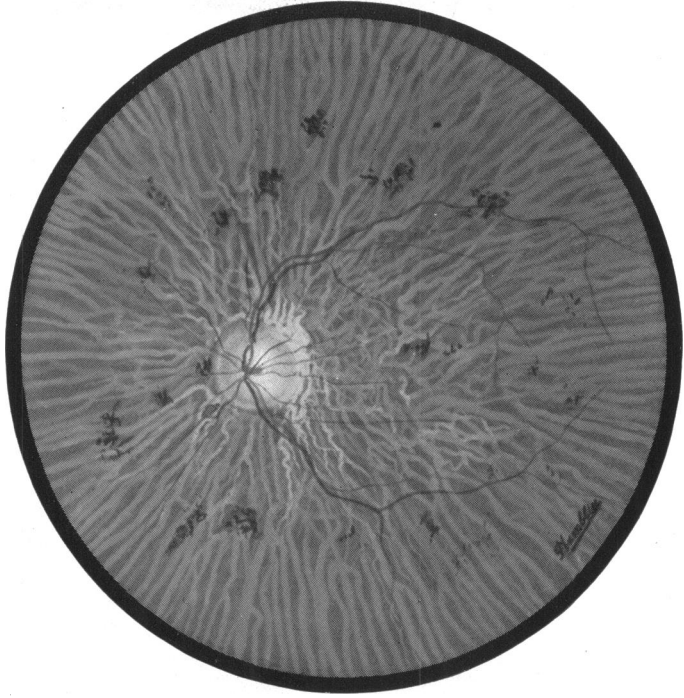


FIG. 17. For legend see Fig. 16.

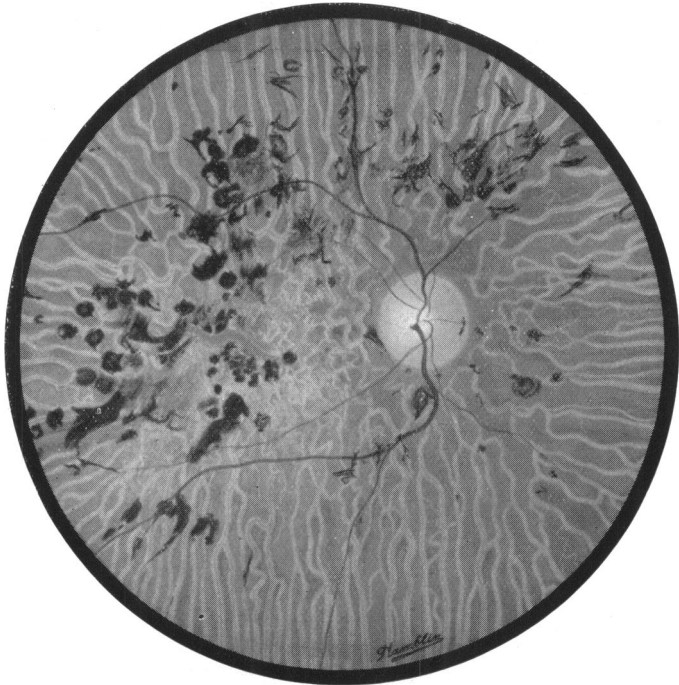


FIG. 18. For legend see Fig. 16.

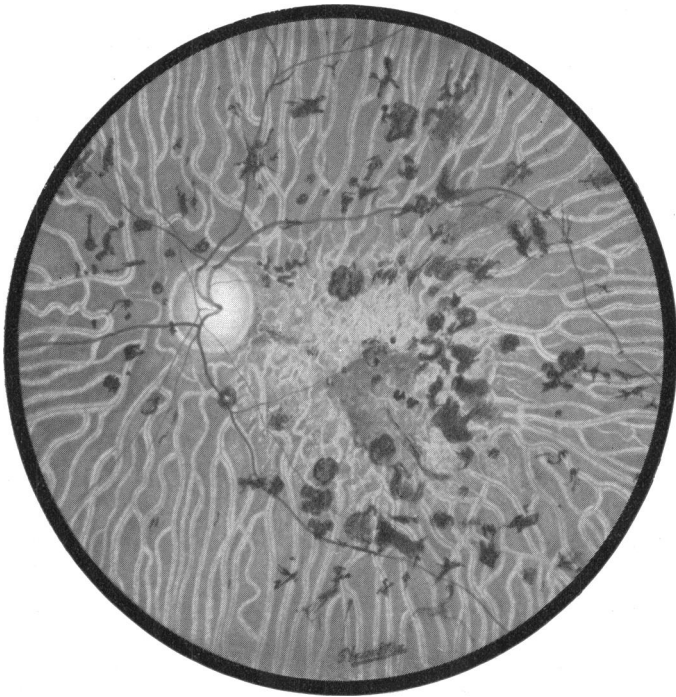


FIG. 19. For legend see Fig. 16.



FIG. 20. RIGHT FUNDUS.

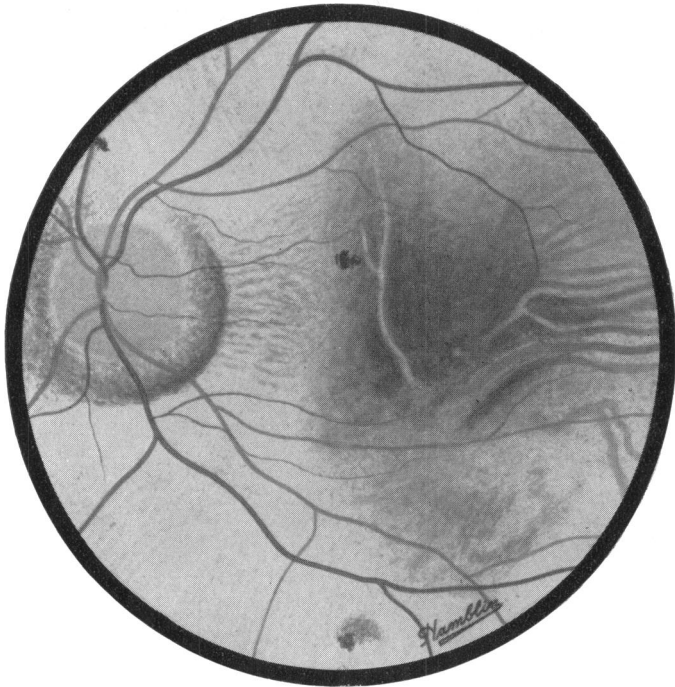
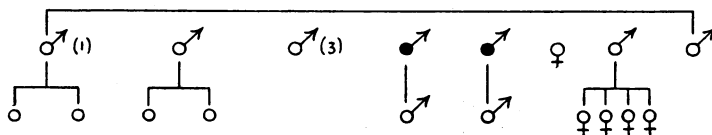


FIG. 21. LEFT FUNDUS.

The subjective symptoms in this patient suggested retinitis pigmentosa, but the ophthalmoscopic changes indicate the disappearance rather than sclerosis of the choroidal vessels. An elder brother shows similar fundus appearances and in an elder sister the lesion is confined to the central areas.

some peripheral extension of sclerōsis of choroidal vessels (Figs. 1 and 2).

Family history : parents had no eye trouble, and there are no antecedents of any significance. No consanguinity. There were eight children. (1) and (3) are dead. Patient is the fifth member of his family. His immediate elder brother (4) has bad sight, "like himself." All other members of the family are reported



not to have any trouble. The only sons of the patient and his affected brother were examined and found to have normal vision and normal fundi. The affected brother Richard R., whose history is given below, is bedridden, suffering from heart trouble. He was seen at his house and a report on his general condition was kindly supplied by Dr. J. S. Cotman and Dr. Claude Wilson.

Richard R., aged 63 years. Vision : 3/60 each eye. Fields full to hand. No night blindness. Like his brother was unaware of any eye trouble till age of 20 years or so. Sight failed progressively till aged about 50 years. The fundi show essentially the same changes as in his brother, except that the lesion is more sharply circumscribed (Figs. 3 and 4). *General condition* : "When first seen some ten months ago, he had cardiac heart failure (breathlessness, cough, pulse rate 160, liver down to umbilicus). Treatment and rest improved the condition, but the man is a chronic invalid. B.P. 190/110. Heart enlarged; peripheral arteries hardened. Always cyanosed and rather dyspnoeic. No cardiac murmur." *Cardiological report* : "Many premature systoles arising in right ventricle and a few in left ventricle. The normal beats in lead II do not suggest serious myocardial damage, though this is not excluded. Some L.V. preponderance (in axis deviation). On screening, heart not enlarged."

(ii) PERIPAPILLARY CHOROIDAL SCLEROSIS. Mrs. E. N., aged 63 years. Complained of sudden loss of vision of the left eye. Vision Rt : $c+1.0=6/5$. Left : 1/60, N.I. The peripheral field is full and there is no night blindness. Ophthalmoscopically both eyes showed peripapillary choroidal sclerōsis and there was a large central choroidal haemorrhage in the left eye (Figs. 5 and 6). The absorption of the choroidal haemorrhage and the extension of the choroidal sclerōsis with the progressive disappearance of the sclerōsed choroidal vessels are illustrated in Figs. 7 and 8, drawn respectively four months and eight months later. General examination revealed no abnormalities (B.P. 165/90).

The family history was negative apart from the fact that a sister, Mrs. B., two years older, was said to have been blind for the past three years, and a sister aged about 50 years is said to have no eye trouble. There was no consanguinity. The only daughter of the patient lives abroad and is said to have no eye trouble. The blind sister lives at Hull and through the kind help of Mr. Stenhouse Stewart it was possible to have her fundi drawn. As can be seen from Figs. 9 and 10 they illustrate a more advanced stage of the same features as seen in Mrs. E. N. There is more actual disappearance of choroidal vessels, the condition approaching in appearance the last drawing of Mrs. E. N. (Fig. 8). A marked feature is the greater degree of pigmentary changes. In neither case is there any marked changes in the retinal vessels.

(iii) CENTRAL AND PERIPAPILLARY CHOROIDAL SCLEROSIS. Gertrude C., aged 58 years, and Edith C., aged 59 years. The fundus appearances, shown in Figs. 11, 12, 13, 14, are essentially similar in the two patients. There is peripapillary choroidal sclerosis, involving the macula and producing heavy pigmentary disturbances. The periphery is clear. Vision: 6/60; the peripheral fields are full and the patients get about quite well. Visual disturbances began in the younger sister fourteen years and in the older ten years ago. The younger sister shows a somewhat raised blood pressure: 175/95; the radial vessels are rather thickened. The elder sister has rheumatoid arthritis and a raised blood pressure: 190/95. The radial arteries are markedly thickened, and exertion brings on shortness of breath. There is no definite heart lesion. The W.R. in both patients is negative.

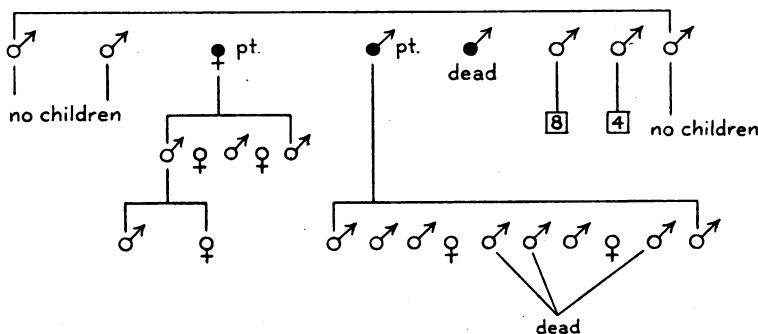
Family history: No consanguinity. The father died at 49 years from "fatty heart," the mother at 64 years from "consumption." There were four children, the patients—the two youngest—are the sole survivors; they are unmarried. An elder sister died at 46 years from "cerebral haemorrhage" and a brother at 41 years from "consumption."

A point of interest is the availability of a fundus drawing at the beginning of the affection in the case of the older sister. She was seen by Mr. F. A. Williamson-Noble in 1926. There was at that time no evidence of choroidal sclerosis and the condition was considered as central choroiditis of possibly tuberculous origin. By courtesy of Mr. Williamson-Noble the fundus drawing is reproduced here (Fig. 15).

(iv) GENERALISED CHOROIDAL SCLEROSIS. (1) Thomas S., aged 60 years, and Mrs. R. L. (a sister), aged 62 years. Vision has been failing since childhood and is now reduced to P.L. There is no definite history of night blindness; in fact, there is a history of sight being better in the evening. Thomas S. is dark haired

and his fundi give a deep colour. Otherwise the appearances are similar to those seen in his sister (Figs. 16, 17, 18, 19), except that the retinal arteries are almost normal in appearance in his case in contrast to the marked narrowing seen in his sister. General investigations, including Wassermann tests: negative.

There are no significant antecedents in the family. The parents were not consanguineous and had good sight. The mother, aged 83 years, is alive. There were 8 children and there is a history of blindness in a younger brother, now dead.



(2) In contrast to this clear cut type of familial choroidal sclerosis, the following case-report illustrates choroidal sclerosis closely simulating retinitis pigmentosa and the end stage is of the type that is erroneously described as choroideremia by some observers.

W. A. C., male, aged 43 years. Parents non-consanguineous. Vision: R. 6/24, L. 6/6. Fields reduced to fixation point. Night blindness. Sight always defective, but became troublesome at about 25 years. The retinal arteries and the discs do not suggest retinitis pigmentosa (Figs. 20 and 21). A brother 11 years older shows the same fundus appearances, except that the arteries are narrow. His vision is 6/9 each eye and the subjective symptoms are so mild that he is barely handicapped. Of particular interest are the ophthalmoscopic appearances in a sister aged 51 years, suggestive of an old central choroiditis. The discs, retinal vessels and periphery are normal. She is not aware of any visual disturbance; her visual acuity is R. 6/6; L. 6/9.

(3) Discussion

Choroidal sclerosis as a clinical entity does not appear to be well established in the literature or in clinical practice. Yet the condition is not particularly uncommon. The older literature does no doubt contain many case reports erroneously interpreted

as the end stage of inflammatory lesions. There are perhaps also records additional to the quoted reports of atypical retinitis pigmentosa which might more properly be regarded as cases of choroidal sclerosis. The review of literature and the personal observations establish choroidal sclerosis as a distinct clinical entity of a familial character. The recognition of the familial character is of value in enabling a study of the different stages of the affection. A family group giving this opportunity has recently been observed by Mr. Frederick Ridley and will be reported in a subsequent paper.

Night blindness and undue contraction of the field was not a feature in any of the patients reported in the present study if the second family showing generalised choroidal sclerosis is excluded—and in these particular patients the fundus appearances suggested atrophy and disappearance of the choroidal circulation rather than sclerosis. In their case the diagnosis of "atypical" retinitis pigmentosa would not be as unwarranted as it would in the other patients.

The distinction between a peripapillary type and a central and peripapillary type of choroidal sclerosis has some justification. Were it not for the previous history, one would be tempted to consider the two patients with central and peripapillary choroidal sclerosis as illustrating a later stage of the affection seen in the two sisters described as showing only peripapillary choroidal sclerosis. Documentary evidence does, however, show that the affection began as a central lesion and that the peripapillary involvement came later.

Just what relationship there exists between these different forms of choroidal sclerosis, is a matter for further study, as are the intermediate stages of these affections and their possible general associations.

The recognition of choroidal sclerosis as a clinical entity raises the problem of the significance of sclerosis of the choroidal vessels in retinitis pigmentosa. The conflicting evidence on the presence of choroidal sclerosis in this affection complicates the issue. The older view that this vascular disturbance was the underlying factor in all cases of retinitis pigmentosa is no longer widely held. A series of observers—Gonin (1903), Stock (1908), Ginsberg (1908), Suganama (1912) and Verhoeff (1930)—have stressed the disproportion between the retinal lesion and the choroidal changes. Ginsberg who emphasised that the cause must be sought in the neuro-epithelium itself, pointed out that the retina may remain intact when there are gross changes in the choroid, as in long-standing glaucoma and nephritis.

The weight of histological evidence is in favour of regarding the neuro-epithelium as the primary seat of the affection, but the

choroidal factor cannot altogether be ignored. Clinically some cases of retinitis pigmentosa show considerable choroidal sclerosis with relatively little retinal involvement, as judged by the pigmentary disturbances and the appearance of the disc. There is, therefore, some validity in the view advocated by Ascher that retinitis pigmentosa shows two forms, one mesodermal or choroidal, and the other ectodermal or retinal. Some genetic support for this distinction has been advanced by Wibaut. How far these types have a distinct genetic difference and different general associations is still questionable. What is of importance is to recognise that if the choroidal type of retinitis pigmentosa is indeed a clinical entity, it differs considerably from the familial cases of generalised choroidal sclerosis to which attention is drawn in this paper. It is possible that the two familial groups of generalised choroidal sclerosis described here may represent these two types—the S. family being an illustration of choroidal sclerosis and the C. family of the choroidal type of retinitis pigmentosa.

The nature of choroidal sclerosis requires elucidation. The present case reports establish its familial character and the fact that the lesion develops in adult life. Presumably the choroidal circulation earlier in life was normal. In so far as the lesion appears to develop in post-natal life in fully differentiated tissue, and is of a hereditary character, it may be regarded as belonging to the group of heredo-degenerations or abiotrophies.

My thanks are due to Mr. F. A. Williamson-Noble for permission to use Fig. 15, to Dr. J. S. Cotman, the late Dr. Claude Wilson and to Mr. Stenhouse Stewart for the trouble they have taken in supplying information on patients under their care.

REFERENCES

(1) CHOROIDAL SCLEROSIS.

- BEDNARSKI.—*Arch. f. Augenheilk.*, Vol. XL, p. 420, 1900.
 CUPERUS.—*Arch. f. Augenheilk.*, Vol. XLVIII, p. 235, 1903.
 DI MARZIO.—*Fundus Oculi*. Rome, 1938. Figs. 194, 195 and 196.
 FRANÇOIS.—*Bull. Soc. belge d'Ophtal.*, Vol. LXXII, p. 74, 1936.
 FROST.—*Atlas Fundus Oculi*. London, 1896. (a) Plate 56. (b) Plate 60. (c) Plate 61.
 GILBERT.—In Schieck and Brückner *Kurzes Handb. d. Ophthal.*, Berlin, Vol. V, p. 133, 1930.
 GUGLIANETTI.—*Arch. di Ottal.*, Vol. XVI, p. 1, 1909; *Ibid.*, Vol. XV, p. 469, 1908.
 HAAB.—*Atlas of Ophthalmoscopy*. English translation. London, 1928. Plate 81.
 HARMAN.—*Trans. Ophthal. Soc. U.K.*, Vol. XXII, p. 156, 1902.
 HOLLOWAY.—*Trans. Coll. Physicians Philadelphia*, Vol. XXXVI, p. 316, 1914.
 HOLLOWAY and FEWELL.—*Trans. Coll. Physicians Philadelphia*, Vol. XLI, p. 285, 1919.
 JAEGER.—*Beiträge zur Pathologie des Auges*. Vienna. First Edition, 1855. (a) Plate 9; (b) plates 5, 6 and 20. Second Edition, 1870. (a) Plate 51; (b) plates 52, 39 and 20.

- KAPUSCINSKI.—*Ber. deut. Ophthal. Gesellsch.*, Vol. L, p. 13, 1934.
 KNAPP.—*Klin. Monatsbl. f. Augenheilk.*, Vol. CDLI, p. 171, 1907.
 LEVINSON.—*Arch. f. Augenheilk.*, Vol. XXXVIII, p. 268, 1899; *Arch. of Ophthal.*, Vol. XXXII, p. 6, 1903
 MORTON.—*Trans. Ophthal. Soc. U.K.*, Vol. V, p. 142, 1885; *Ibid.*, Vol. XIII, p. 282, 1893.
 NETTLESHIP.—*Trans. Ophthal. Soc. U.K.*, Vol. IV, p. 165, 1884.
 OELLER.—*Ophthalmoskopischer Atlas*. Wiesbaden, 1899. Plates D.14, D.15 and D.7.
 ——— *Atlas seltener ophthalmoskopischer Befunde*. Wiesbaden, 1905. Plate D.1.
 PILLAT.—*Amer. Jl. of Ophthal.*, Vol. XIII, p. 1, 1930.
 PÖLLOT.—*Arch. f. Ophthal.*, Vol. LXXX, p. 379, 1911.
 RETZE.—*Beiträge zur Augenheilk.*, Bd. V, Heft 47, S. 94, 1901.
 THOMPSON.—*Trans. Ophthal. Soc. U.K.*, Vol. XXV, p. 118, 1905; *Ibid.*, Vol. XXI, p. 66, 1901.
 WEISS.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LII, p. 142, 1914.
 WILMER.—*Atlas Fundi Oculi*. New York, 1934. Plate 82.

(2) OTHER REFERENCES.

- ASCHER.—*Ber. deut. Ophthal. Gesellsch.*, Vol. XLIX, p. 547, 1932; *Arch. f. Augenheilk.*, Vol. CVI, p. 585, 1932.
 GINSBERG.—*Klin. Monatsbl. f. Augenheilk.*, Vol. XLVI, p. 1, 1908.
 GONIN.—*Ann. d'Ocul.*, Vol. CXXIX, p. 24, 1903.
 SORSBY, AVERY and COCKAYNE.—*Quart. Jl. Med.*, N.S.; Vol. VIII, p. 51, 1939.
 STOCK.—*Klin. Monatsbl. f. Augenheilk.*, Vol. XLVI, p. 225, 1908.
 SUGANUMA.—*Ibid.*, Vol. L, p. 178, 1912.
 VERHOEFF.—*Arch. of Ophthal.*, Vol. LXII, p. 392, 1931.
 WIBAUT.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LXXXVII, p. 299, 1931.

A CASE OF ALEUKAEMIC LYMPHOSIS INVOLVING THE UPPER LIDS. WITH PATHOLOGICAL FINDINGS*

BY

FREDERICK T. TOOKE

MONTREAL, CANADA

(FROM THE DEPARTMENT OF OPHTHALMOLOGY ROYAL VICTORIA
HOSPITAL, MONTREAL, CANADA.)

THE globe and its appendages are occasionally the site of abnormal proliferation of the lymphoid tissues. In very rare cases these occur as local tumour growths, either of benign lymphomatous, or of malignant lymphosarcomatous character. In the vast majority of cases, however, the lesion of this region is merely part of a systemic disease which sooner or later comes to involve more or less of the lymphoid tissue of the body. Consequently, any of the various forms of systemic proliferation of lymphoid tissues may express itself in this region, and hence a great variety

* Read before the American Ophthalmological Society, San Francisco, June 8, 1938.