THE BRITISH JOURNAL
OF
OPHTHALMOLOGY

MAY, 1922

COMMUNICATIONS

AN UNUSUAL CASE OF RENAL RETINITIS

BY

R. FOSTER MOORE, O.B.E., F.R.C.S.

WITH PATHOLOGICAL REPORTS BY

DR. GEOFFREY EVANS, AND HUMPHREY NEAME, F.R.C.S.

ASSIST. DIRECTOR, MEDICAL UNIT,
ST. BARTHOLOMEW'S HOSPITAL

PATHOLOGIST AND CURATOR TO THE
MOORFIELDS EYE HOSPITAL

THE following case of renal disease with retinitis seems to be worth publishing in considerable detail, on account of its unusually interesting course and the completeness with which various examinations have been possible.

Briefly, it is the case of a man, who, at the age of 45, was ill for the first time in his life, and who then sought advice for failing sight which was found to be due to renal retinitis. The retinal changes were so severe as to produce detachment of the retina in each eye.

In the course of months the retinitis subsided, the detachments disappeared, and he was exhibited at the Congress of the Ophthalmological Society in 1915 (*Trans. Ophthalm. Soc., U.K., 1915, XXXV, 159*), with a coloured drawing of his fundi at that time, showing a retinitis which was well advanced towards subsidence, with a partial atrophy of the disc and a complete absence of retinal detachment.

From this time on he continued at his work at a " Small Arms " factory, at times doing seven days a week of 80 hours, and seven years later he was admitted to St. Bartholomew's Hospital where

he died. Thus, in addition to a closely followed clinical course and repeated ophthalmological examinations, a complete examination of the body was made, and a complete histological examination of both eyes.

The following details are abstracted from my full notes made at each visit.

The dates refer to the period that had elapsed since he first came under observation.

W.C., aged 45, first attended the Moorfields Eye Hospital on May 2, 1913, under Mr. Herbert Fisher, on account of failing sight. He stated that until six weeks before this date he never had had a day's illness in his life.

Five weeks ago he noticed that his sight was defective, and about the same time had headaches, nausea, and pain in the back.

He said he had never had syphilis or scarlet fever.

His urine was of a specific gravity of 1.014, and contained much albumen.

His systolic blood pressure was 215 mm., he had no oedema. He was examined by Dr. James Taylor, who reported: "Heart hypertrophied and dilated, probably sclerotic kidneys."

Mr. Fisher, knowing of my interest in him, was good enough to transfer him to my care.

After eleven days. The eyes were normal except for the fundus changes, his visual acuity was R. 6/36, and L. less than 6/60.

Ophthalmoscopic examination

Right eye. The disc was swollen and the edges much obscured; there was copious retinal exudate over the central region; it was not arranged in the form of a star figure; the veins were engorged and were much cut into where they were crossed by the arteries.

Left eye. There was much massive exudate which obscured the vessels in places, the changes did not differ essentially from those present in the right eye; in each retina a number of haemorrhages were present.

There was a rather small retinal detachment below in each eye.

After three weeks. The veins were very dark, distended, and tortuous; there was more exudate which now tended to form a coarse-textured star figure and was becoming conglomerate.

The blood pressure was 205 mm., the urine was unchanged; the retinal detachments were present as before.

After five weeks. At this date, before examining him myself, I asked Mr. Fisher to see him, who sent a note saying, "I agree to the detachment in the right eye, but I cannot see one in the left," and on examination it was evident that the detachment had gone from the left eye.

After eight weeks. There was now no detachment in either eye, but otherwise one could not appreciate any essential difference since last time.

After four months. R.V. 6/36, L.V. 2/60.

His general health had improved, his blood pressure was 235 mm. The fundus changes were still profuse, but now for the first time one could be sure that the veins were less engorged and were becoming craggy in their contours, the first sign of subsidence of the retinitis.

FIG. 1.—The retina near the disc showing:—

- (A) Two small masses of retinal exudate in Henle's layer.
- (B) The base and (c) the apex of the pigment process in the rosette.
- (D) Choroid.
- (E) Pigment epithelium.
- (F) Sclerotic.

There were no detachments to be seen, and there was no recurrence of them up to the time of death (see Mr. Neame's report).

After five months. There were now marked evidences of subsidence of the retinitis. All the vessels, both arteries and veins, were much reduced in size, the veins had white lines along them for a considerable distance from the disc, and in one or two places the arteries showed "pipe-stem sheathing."

The discs were becoming pale and hazy edged and were taking on the characters of a secondary optic atrophy.

After six months. For the first time at this date the small

angular pigment spots which have been figured by Liebreich (Atlas Tab. ix, Fig. 1 and 2), and described by Nettleship (*Trans. Ophthalm. Soc. U.K.*, 1899, XIX, 63) were noticed towards the periphery.

The veins now showed well the irregular craggy bends instead of full curves, such as one sees when greatly distended veins later become reduced in size.

After seven months. He is now extraordinarily improved in health.

FIG. 2.—Showing (A) the main part of the pigment process in the form of a loop.
Outlying pigment cells lie close to the anterior surface of the retina.

Right eye. All the fundus changes are retrogressing and the exudate is becoming powdery in appearance; there are still a few haemorrhages and patches of more solid exudate.

Left eye. In this eye all the exudate is of the fine powdery type, there are no solid masses; the pigment spots are unchanged, they are more circular than those depicted by Liebreich.

After eight and a half months. R.V. 6/24, L.V. 3/60.

His health is improving, there is a haze only of albumen in the urine, the blood pressure is 220 mm.

The exudate is less, there are three or four haemorrhages only.

After ten months. R.V. 6/36, L.V. 6/60. Blood pressure 210 mm.

After eleven months. Right eye. The disc is pale and hazy

edged as before, and the vessels are very fine, the rest of the fundus in general is normal, except for the fine powdery exudate which is much less than in the drawing previously made by Head.

Left eye. The disc is pale and the edges are indefinite, but are becoming sharper; I can find one haemorrhage only.

There is still a very little powdery exudate within about a disc's breadth of the yellow spot, and the rest of the fundus is nearly normal.

FIG. 3.—A vertical sagittal section of the posterior half of the left globe showing complete absence of detachment of the retina.

(A) Pigment mass.

Section between the plane of those shown in Figs. 1 and 2.

There is the very faintest trace of albumen in the urine.

After fifteen and a half months. He is still doing well, the vision is 6/18 and 6/60, the blood pressure is now 190 mm., and there is not a trace of albumen in the urine.

After nineteen months. The only change in either fundus now, besides the signs of secondary atrophy and the very attenuated vessels, is a very little exudate.

After seven years. Here is a gap of five and a half years during which I did not see him. He was, however, working overtime,

for the greater part of this period in a " Small Arms " factory; I was greatly surprised when one morning he came up to my desk, I had expected he was dead years ago.

He expressed himself as being very well except for a cough. His vision was 6/18 and counts fingers, his blood pressure was 265 mm., and he had a heavy cloud of albumen in his urine.

The fundi were but little altered since last he was seen, there was not a trace of exudate, there were no haemorrhages; for the rest the discs had the characters of secondary atrophy, and the vessels, especially the arteries, were very attenuated. A few weeks later he was taken ill, and was admitted to St. Bartholomew's Hospital under the care of Dr. J. H. Drysdale; he died a few weeks later in uraemia.

I last examined him ophthalmoscopically the day before he died. A complete post mortem examination and a histological examination of the kidneys was made by Dr. Geoffrey Evans whose report is attached.

I removed both eyes entire. They were placed in Zenker's fluid and were thoroughly examined by Mr. Humphrey Neame, Pathologist and Curator to Moorfields Eye Hospital, whose report is also attached.

Remarks

It is not often that one is so fortunate as to be able to trace a case of this nature from within six weeks of the first symptoms of illness, up to the time of death, and then to obtain so complete an examination. The features that seem especially worthy of amplification and comment are: the length of time of survival after the development of renal retinitis, the retinal detachments and their disappearance, the subsidence of the retinitis as watched with the ophthalmoscope, and the pathological findings both in the body generally and in the eyes.

The long accepted formula is undoubtedly true, that few patients live so long as two years after the discovery of renal retinitis. Thus Belt (*Jl. Amer. Med. Assoc.*, 1895, XXV, 735) found that of 419 patients, 6 per cent. lived longer than two years and 65 per cent. died within a year, and many other statistics are available which are in general agreement with this.

There are one or two groups of cases in which it is notorious that the prognosis is much less grave, as in the nephritis of pregnancy, and probably in " trench nephritis."

This patient, however, seems to have been a case of chronic mixed nephritis, and his heightened blood pressure when first seen, 215 mm., seems to indicate a nephritis of some long standing.

The retinal detachments. Retinal detachments in severe renal

retinitis are by no means rare if they are expressly looked for up to the time of death: I was able to find thirteen cases in two and a half years at St. Bartholomew's Hospital (*Royal Lond. Ophthalm. Hosp., Moorfields, Reports*, 1916, Vol. XX, p. 262).

They occur most frequently in cases where there is much retinal exudate, and are especially common in pregnancy cases.

They are, I believe, always bilateral, and involve the lower part of the retina, and if the patient survives they are probably always recovered from.

I have seen the sub-retinal fluid completely absorbed within eight days of its discovery.

Apart from pregnancy cases the prognosis is especially bad, for detachment occurs almost exclusively in those cases in which the retinal changes are severe.

Subsidence of the retinitis. Seeing that retinitis precedes death in nephritis for varying periods, but in general for a short time only, there are comparatively few cases in which it is possible to trace the gradual disappearance of the retinitis to a point at which it may be said to have completely subsided.

Nevertheless retinitis runs a definite course; it increases in severity till an acme is reached, and afterwards as subsidence sets in, retrogressive changes take place until all signs of active disease are gone, and a varying degree of secondary retinal and optic nerve atrophy is left.

During the progressive stage, individual patches of exudate and haemorrhage become absorbed in a few weeks, but fresh areas appear and so the picture is maintained. Discrete spots of exudate may become confluent and so give rise to massive areas, but there comes a time if the patient survives, when absorption is in excess of fresh formation, till no fresh changes occur; thus it may happen at the time of death that retinitis is in its early stage, or at its acme, or well advanced towards subsidence.

The first sign of subsidence is a reduction of the turgid condition of the vessels till they ultimately are smaller than normal instead of being engorged. The veins make short craggy bends instead of full curves, the appearance suggests that they have been so greatly, and so long distended that their resilience is impaired, and so they have become permanently stretched; they often have white lines along them.

The arteries become greatly reduced in calibre and may show "pipe-stem sheathing" which, however, afterwards disappears in spite of its solid look.

The exudate becomes less in amount, and it is very characteristic that its remains are of a fine white dusty or powdery appearance, and in this condition it remains for long.

The pigment spots already described may be found in scanty numbers, chiefly some distance away from the central regions. I have seen one of these spots which seemed to develop out from a preceding area of white exudate.

In the present patient at the end of a year the signs of active changes had largely gone, and at the end of seven and a half years, that is one day before his death, the signs of a well-marked secondary atrophy of the nerve and retina were present, there was not a trace of exudate and no haemorrhages: it was clear that the retinitis as such had long ago become completely obsolescent; it was remarkable how little histological change was to be found in the retina (see Mr. Neame's report). There were no signs of the previous retinal detachments (Fig. 3).

It is a point of considerable interest to enquire whether a retinitis, once subsided, can recur should the nephritis become recrudescent.

There is some evidence to show that an eye which has been the subject of serious pathological processes of any kind, is, by this means, protected against the occurrence of retinitis in nephritis.

Thus Nettleship reports a case in which one eye only was affected, the other being blind as a result of thrombosis of the central retinal artery, and a unilateral case in which the unaffected eye was highly myopic (*Royal Lond. Ophthalm. Hosp., Moorfields, Reports*, 1903, XV, 340). I have seen a similar case.

With regard to the present case, very few patients die in uraemia without some fundus changes, and I feel no doubt that he would not have died in the manner he did without developing retinitis, had not his eyes been protected against its occurrence as a consequence of the atrophy following the previous disease.

Dr. Geoffrey Evans's Report

W.C. died 15/11/20, post-mortem 16/11/20. Dr. Drysdale. Post-mortem book 1920, Vol. XLVII, p. 212.

P.M. Diagnosis.—Uraemia; chronic mixed nephritis; arterio-sclerosis; nutmeg liver; oedema and infarction of the lungs.

External appearances.—Wasted and pale.

Head and neck.—Brain normal; arteries at base showed slight patchy nodular sclerosis in basilar and vertebral arteries, and a similar lesion was present to a more marked extent in the branches of the middle cerebral arteries.

Thorax.—Lungs engorged and oedematous; patchy pleurisy over both lungs and numerous adhesions, old and recent. Massive oedema of left lower lobe. Multiple infarction of both upper lobes, areas chiefly superficial and size varying between a walnut and a pea. Brown induration marked.

Heart.—No pericarditis; petechial epicardial haemorrhages. Mitral orifice admits three fingers; marked hypertrophy and dilatation of left ventricle; dilatation and slight hypertrophy of right. Patchy sclerosis of both mitral and tricuspid valves. Aortic and pulmonary valves normal. Heart muscle firm, not fatty.

Aorta and vessels.—Ascending aorta normal except for plaques of sclerosis in the sinus of Valsalva. Sclerosis of transverse arch increasing in descending thoracic aorta to a coalescing sclerosis of advanced degree showing hyaline areas of degeneration (slight), fatty areas of degeneration (marked) without obvious calcification. All vessels showed medial hypertrophy.

Abdomen.—Peritoneum normal. Stomach and intestines *nil ad rem*.

Liver.—Nutmeg.

Spleen.—Firm and fibrous, tiny arteries stand out prominently.

Suprarenals.—Very marked hypertrophy.

Kidneys.—Right showed a depressed scar opposite the pelvis and other small scars on the surface. Both had a mottled colour, purple, with paler areas. The capsule was adherent to the perirenal tissues and adherent to the renal cortex; it was thickened and split on stripping leaving a coarsely granular surface in which the depressions were a deep purplish colour and the granules pale. On section firm; the cut surface—cortex reduced, confused structure, pale in comparison with the pyramids. Pelvis increased in size and congested. Vessels thickened and showed obvious sclerosis.

Mr. Humphrey Neame's Report

W.C. Post-mortem November 16, 1920.

Macroscopic examination. The eyeballs were both fixed in Zenker's fluid. The *right* was divided horizontally immediately below the disc, but including in the lower half the greater part of the stump of the optic nerve apart from the disc. The upper half is embedded in celloidin, the lower half is in 4 per cent. formaldehyde.

The lower half of the right eye shows a cornea of normal appearance, opaque from fixation, and anterior chamber measuring 2.5 mm. between the pupil margin and the posterior corneal surface immediately opposite to it. The angles of the anterior chamber are well open. The iris and ciliary body appear normal. The lens is of normal appearance apart from slight tearing of the posterior capsule. The vitreous cavity contains a veil-like grey-white substance behind the lens and ciliary body, but behind this narrow zone the main part of the space is perfectly clear.

The retina is *in situ*, and there is nothing to suggest that there had ever been any retinal detachment. The choroid is *in situ*, and there is no apparent exudate external to this structure, or between it and the retina. The sclerotic is of normal appearance.

The left globe is also in two halves, divided in the equatorial plane or slightly anterior to this. The anterior half viewed from behind shows lens, ciliary processes, anterior part of retina and choroid *in situ* and of normal gross form and structure. The posterior half is embedded in cellodine. Division of the latter parallel to the sagittal plane, between disc and macula, shows, on the cut surface, sclerotic, choroid and retina of usual appearance and with no sign of detachment or of exudate or coagulum between the layers.

The macroscopic examination of the right and left eyeballs, therefore, shows no trace of retinal detachment.

Microscopic examination. Sections stained with Ehrlich's haematoxylin and with eosin, or with Weigert's iron haematoxylin and van Gieson stain.

Right eye. The cornea, its epithelium, membranes of Bowman and Descemet and its endothelium appear normal. The canal of Schlemm is patent, the anterior chamber of moderate depth and its angles are well open. The iris, ciliary body and lens show normal structure. Nothing is seen in the vitreous cavity. The retinal pattern shows slight changes. There is some cystic degeneration of the anterior part, and a granular and fragmented appearance of the rod and cone layers in parts of the sections. In some places this appears to be due to oblique section of this layer. The superior nasal and temporal retinal arteries, cut transversely at the disc, show unusually thick walls. Thorough examination of all the sections cut, reveals no trace of exudate in the retina of this eye. The retina is *in situ* except for the very slight separation that so frequently takes place during the process of dehydration and embedding. The choroid shows a few colloid bodies on the membrane of Bruch. Many of the vessels of this layer are unusually thick walled, otherwise the choroidal structure is normal. The sclera and conjunctiva are normal.

Left eye. Posterior half. In general the microscopic appearance of the structures in the posterior half of this eye is similar to that of the right. There is no pathological detachment of the retina.

A few of the sections which pass vertically through the papilla and optic nerve show, in some one, in others two, globules of exudate in Henle's layer. They are situated at about one and a half disc diameters from the edge of the papilla. These globules lie in small spaces within this layer, as though either they had become shrunken in preparation or they had undergone a process

of partial absorption and that the surrounding fibres of Henle's layer had not, so far, regained their normal position, or had disappeared. The globules show a mauve colour from retention of both haematoxylin and eosin. They have a finely granular and fibrillated structure.

A small portion of the posterior region of the globe, including the nasal margin of the disc was re-embedded by Jordan's photoxylin-paraffin method, and of this a series of particularly thin sections was thus obtained. These sections show in places a condition of the rod and cone layer similar to that already described for the right eye, but to a slighter degree. There is also a vacuolar condition of the outer reticular layer. In the latter, formed by the fibres of this layer, there are many irregular polygonal spaces with a diameter equal to that of from two to four of the nuclei of the outer nuclear layer.

A peculiar pigmentary change is to be observed in the portion of the retina which lies between the disc and the globules of exudate already mentioned. In a series of consecutive celloidin sections there can be traced a mass of pigment cells which arises from the pigment epithelium layer and extends forwards apparently to push the adjacent retinal layers in front of it and to invaginate them into the anterior layers. Where this pigment cell mass arises from the epithelial layer the latter is in most of the sections continuous, and in close apposition with the choroid. The mass of pigment appears to be a finger-like process which describes an arc, of which the extremity comes to lie at about the middle of the thickness of the retina, but nearer to the disc than its base. The retina in this area is almost double the thickness of that in the surrounding region. The pigment process, at the cross-section near its extremity is surrounded by a ring of rod and cone nuclei, external limiting membrane and rods and cones, in the form of a large rosette. The rod and cone ring is innermost, and immediately surrounds the pigment. This seems to indicate that, as the pigment cells proliferated and grew forwards they pushed the layers of the retina in front of them. Much pigment proliferation has occurred in the surrounding retina, so that there are many isolated pigment masses. Some of these pigment masses have passed forwards into the nerve-fibre layer, and one is within about 20 micro-millimetres of the anterior limiting membrane. There are many small darkly-stained round nuclei in this affected area, in all layers, and a few elsewhere in the retina. van Gieson's fuchsin-picric-acid stain fails to reveal any collagen fibres in this area of the retina, but the Weigert iron-haematoxylin displays very clearly the presence of the small round nuclei above mentioned.

It is well recognised that in inflammatory conditions of the retina

and choroid, due to the action of micro-organisms or their toxins, there is an overgrowth of the pigment epithelium and a migration of masses of these cells of various sizes into the retina. But the outgrowth of a pigment process of considerable length, which carries before it the layers of the retina into the middle of that structure, seems new. Furthermore, in this case, there is no evidence of an infection with micro-organisms, and the choroid is unaffected, except for a slight degree of colloid degeneration of the membrane of Bruch—of the inner or cuticular lamella of the lamina vitrea. The fact that, in retinal detachment, the separation takes place between the retina and the pigment epithelium layer, would seem to discount the remote possibility that this microscopic appearance resulted from an extreme dimpling of the retina with its pigment epithelium from without, on restoration to its position against the choroid.

Summary. Macroscopic examination reveals no trace of retinal detachment in either eye. Microscopic examination discloses no retinal detachment in either, but shows a slight amount of exudate in Henle's layer in the left eye, and also a peculiar localized pigment proliferation in the same locality of the retina.

A CASE OF TUBERCULOUS IRIDOCYCLITIS AND PARENCHYMATOUS KERATITIS OF THE LEFT EYE, ASSOCIATED WITH TUBERCULOSIS OF THE CON- JUNCTIVA OF THE RIGHT EYE, AND TUBERCULOUS LYMPHADENITIS*

BY

HUMPHREY NEAME, F.R.C.S.

LONDON

HERBERT DYKE, aged 19 years, came under the care of Mr. E. Treacher Collins at the Royal London Ophthalmic Hospital in June, 1921. The patient's occupation was that of a boot and shoe stock-keeper. He was sent to this hospital from the Victoria Park Hospital on account of loss of sight in the left eye.

His history is as follows: Dyke stated that his general health depreciated during and after a fourteen days' holiday at Southend in August, 1920. He had cycled there from London, and on his arrival felt very tired. Throughout his stay there he felt unable to exert himself. After his return to work in London, one day while stooping down he felt something "go" in his left eye, and then saw "black spots and a flash" with this eye. From that

* Shown at the meeting of the Ophthalmic Section of the Royal Society of Medicine, on Friday, November 11, 1921.