ROLE OF A DISTINCTIVE CHOROIDO-RETINAL LESION IN THE PATHOGENESIS OF RETINAL HOLE

A CLINICAL AND PATHOLOGICAL REPORT*†

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THE association of retinal holes with retinal detachment was described shortly after the use of the ophthalmoscope began (Coccius, 1853).

Since the work of Gonin (1931) it has been generally accepted that the retinal hole is the immediate and causative precursor of the detachment.

Hole formation is not the primary lesion but results from degeneration, usually cystoid in type, or from atrophy of the retina (Zeeman, 1912; Leber, 1916; Gonin, 1934; Vogt, 1936a, b). Possibly a disturbance of the vitreous also precedes hole formation, local fibrillar attachments of the vitreous being capable of disrupting the remaining fine inner layer of the retina (Leber, 1882; Nordenson, 1887; Deutschmann, 1895, 1899; Addario, 1904; Gonin, 1904; Lister, 1924; Rötth, 1933).

The retinal degeneration or atrophy and the local vitreous changes are in turn probably not primary changes. The retinal condition depends upon the nutritional supply which has a double source—the chorio-capillaries and the retinal capillaries. Various factors may affect the nutritional supply. Myopia or perhaps senility may affect the chorio-capillaries (Hanssen, 1919) and unknown factors may cause a disturbance of the fine retinal vessels and so cause retinal degeneration (Iwanoff, 1864; Greeff, 1900; Best, 1904; Zeeman, 1912; Leber, 1916; Rahnenführer, 1916; Bruno, 1936; Vogt, 1936a, b). A choroido-retinitis may lead to obstruction of the capillaries of both choroid and retina (Elschnig, 1914; Gonin, 1923; Vogt, 1929; Arruga, 1931; Duke-Elder, 1940). In the latter connexion a distinctive choroido-retinopathy has been described as frequently present in the neighbourhood of retinal holes (Michaelson, 1955). It is characterized by local pigment disturbance of the choroid and by phlebo-sclerosis in the overlying retina, the choroidopathy preceding the visible retinal changes.

The present report is concerned with a clinical description of further cases in which distinctive choroido-retinopathy was present in the region of a retinal hole. In one case it was possible to examine the eye histologically.

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Clinical Descriptions

Case 1, a married woman aged 45 years, suffered from detachment of the retina. The left eye, which had a myopia of 8 dioptries, is shown in Fig. 1. Close to the retinal hole pigmentary changes and sclerosis of the retinal vessels were seen about 5 mm. from the ora serrata.

FIG. 1.—Case 1, left eye, showing in neighbourhood of retinal hole pigmentary changes and sclerosis of retinal vessels about 5 mm. from the ora serrata.

Case 2, a married woman aged 49 years, had had a retinal detachment in the right eye on March 29, 1955. The upper temporal quadrant of the left eye is shown in Fig. 2, as it was observed during a control visit to the clinic on January 3, 1954. This appearance had developed since 1953. About 5 mm. from the ora serrata was a choroidopathy

FIG. 2.—Case 2, upper temporal quadrant of left eye as on January 3, 1954. About 5 mm. from the ora serrata appears an area of choroidopathy comprising four discrete pigmentary changes, close to each of which the local vein is sclerosed. No hole was then observed despite careful examination.

consisting of four discrete pigmentary changes, close to each of which the local vein showed sheathing and sclerosis. No retinal hole was then observed. This case was reported at the XVII International Congress of Ophthalmology (Michaelson, 1954), when it was stated that the case was being kept under observation. By May 30, 1955, the changes had developed which are noted in Fig. 3. The choroidopathy and the vascular sclerosis had markedly increased and a small retinal hole had developed.

FIG. 3.—Case 2 (cf. Fig. 2), upper temporal quadrant of left eye as on May 30, 1955. The choroidopathy and vascular sclerosis have markedly increased and a small retinal hole has developed.

Case 3, a married woman aged 50 years, suffered from retinal detachment. Close to the retinal hole which was about 5 mm. from the ora serrata, pigmented choroidopathy and phlebo-sclerosis were seen (Fig. 4).

FIG. 4.—Case 3, pigmented choroidopathy and phlebo-sclerosis close to retinal hole, which is about 5 mm. from the ora serrata.

Clinico-Pathological Report

Case 4, a man aged 39 years, who complained of defective vision, was found to have a detachment of the retina in the lower temporal quadrant of the right globe. A minute retinal hole was found in the flat portion of the retina in the 10 o'clock meridian about 5 mm. from the ora serrata. In the neighbourhood of this hole were small areas of pigmentary disturbance and sheathing of a retinal vein (Fig. 5). The visual acuity of the right eye was 5/5 (letters) and the refraction +1 D sph.; the left eye was normal in all respects and had a visual acuity of 5/5.

FIG. 5.—Case 4, enlargement of retinal hole area noted in 10 o'clock meridian.

As there was no apparent connexion between the retinal hole and the retinal detachment observed in the right eye, and as a most definite shadow was found on transillumination of the detached area, enucleation of the globe was performed. The globe was fixed in formalin and thereafter the fundus was examined by means of the slit-lamp after removal of the cornea and lens. Gentle wiping away of the vitreous facilitated the observation of the fundus. It was noted that the vitreous was adherent to the retinal hole area observed clinically at 10 o'clock. The slit-lamp appearance (Fig. 6) shows a

FIG. 6.—Case 4, slit-lamp appearance of retinal hole area. A row of minute holes extends from the single one noted clinically; the phlebo-sclerosis is more extensive than can be observed clinically.

row of minute holes extending from the single one seen macroscopically; the phlebo-sclerosis is more extensive than could be observed clinically. The portion of the globe containing the detached portion of the retina was embedded separately. Sections confirmed the diagnosis of a choroidal sarcoma. The portion of the retina and choroid at the site of the hole area was removed by means of a 4-mm. trephine. Unfortunately the retina and choroid separated from each other and had to be embedded separately. Before embedding the retina, block examination was carried out using a microscope. From this examination a scale drawing was made in order to make certain the exact localization of the changes noted with the help of the ophthalmoscope and slit-lamp. With the help of the scale-drawing and by suitably orientating the retinal block, it was possible to be certain that, for example, section No. 340 was in the hole area.

Histological Examination:

(1) A retinal hole was situated in a local area of extreme retinal atrophy (Figs 7 and 8).

FIG. 7.—Case 4, area of extreme retinal atrophy surrounding retinal hole ($\times 100$).

FIG. 8.—Case 4, complete absence of tissue constituting retinal hole ($\times 80$).

The meeting of the inner and outer nuclear layers at the edge of the hole is shown in Fig. 9. This appearance obviates any possibility that the hole may have been an artefact.

(2) Hyaline changes in the vessel had been observed clinically (Fig. 10), but differential stainings of these sections were not obtained.

FIG. 9.—Case 4, edge of hole illustrated in Fig. 8. The meeting of the inner and outer nuclear layers can be well seen, and obviates the possibility that the hole may have been an artefact ($\times 460$).

FIG. 10.—Case 4, sclerosis of retinal vessel observed clinically (Fig. 1). Haematoxylin and eosin ($\times 460$).

(3) A fibrillar disturbance of the vitreous overlay the area of retinal atrophy. Because of the attachment of the disturbed vitreous to the retina, the inner layer of the latter is apparently pulled away from the remainder of the retina (Fig. 11).

FIG. 11.—Case 4, fibrillar disturbance of vitreous in area close to retinal hole. The attachment between the retina and the disturbed vitreous has apparently caused a splitting of the retinal tissue ($\times 460$).

FIG. 12.—Case 4, atrophy of overlying choroid, the vessels being almost completely obliterated. The torn portion of the choroid probably represents local adhesion to the retina ($\times 100$).

(4) Pigment migration investing retinal vessels.

(5) Atrophy of the overlying choroid in which the vessels are no longer to be seen. The torn portion of the choroid probably represents local adhesion of the choroid to the retina (Fig. 12).

Discussion

To the clinical description of the distinctive choroido-retinopathy noted in the neighbourhood of some retinal holes (Michaelson, 1954) the above histological report adds the following facts:

- (i) The local choroid is atrophic and its vessels almost obliterated;
- (ii) Some of the pigment noted ophthalmoscopically is seen to be intraretinal;
- (iii) The sheathing of the local vessels is due to a hyaline degeneration;
- (iv) There is fibrillar disturbance of the local vitreous;
- (v) More holes may be present than are clinically noted.

The occurrence of a retinal hole in an eye removed because of choroidal sarcoma seems inexplicable. That it was fortuitous was clear from the relative situations of the hole and the detachment caused by the tumour. The histological findings of this case support the clinical findings and suggest that, in the pathogenesis of certain retinal holes, an atrophic pigmented choroidopathy is associated with a local retinal phlebo-sclerosis and a fibrillar disturbance of the adjacent vitreous. The former give rise to an atrophy of the retinal tissues which is possibly torn by the pull of the changed and adherent vitreous fibrils. The pre-requisites for hole formation diversely postulated or emphasized by different authors on theoretical, clinical, and histological grounds are apparently supplied collectively by a single distinctive choroido-retinopathic process. The process would appear to meet the two pre-requisites for degeneration and atrophy of the entire thickness of the retina—closure of the local chorio-capillaries and closure of the local retinal vessels. At the same time this process would appear to be accompanied by degeneration of the local vitreous.

The cause of the process may be a general disturbance not confined to the eye. A report on this will be given in a later publication. In this connexion should be quoted the words of Duke-Elder (1940):

The essential factor in the aetiology (of hole formation) is the preceding inflammation or degeneration which rendered possible its occurrence; it is the process which gave rise to this which is the true cause of the condition and should receive primary consideration in treatment.

Summary

(1) Clinical descriptions are given of a distinctive choroido-retinopathy preceding the development of retinal hole in cases of retinal detachment.

(2) A histological description is given of a lesion of this kind which had been noted and sketched clinically.

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