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COMMUNICATIONS

CONGENITAL VASCULAR VEILS IN THE VITREOUS

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THE following three cases (six eyes) were discovered by one of us (A. McR.) in the course of routine examination. The patients made no complaint other than the need for a refraction test. In all there was relative amblyopia, 6/12 being the maximum acuity obtainable by correction. The patients' ages were 17, 20 and 21 respectively and there was no history of ocular discomfort or inflammation at any time. All three were males.

The first example (described as Case 3) was seen in 1934. The second (Case 1), seen in 1937, volunteered the information that his brother (Case 2) had also defective sight. He was therefore examined and found to have a similar condition. The similarity of all three cases and the presence of the abnormality in both eyes of two brothers, who also resemble each other strongly in complexion and appearance, place the condition with fair certainty as a developmental one.

In many respects the findings appear to link up with those described in cases of congenital retinal fold on the one hand and with persistent hyaloid artery on the other, and offer evidence in

favour of the theory that congenital retinal fold is due to an abnormality of development of the vitreous.

It seems justifiable to consider these cases as belonging to a distinct group which we have called "congenital vascular veils in the vitreous."

Case 1.—John G., aged 17 years.

Right eye, 6/36 Hm. +3.0 D.	} Vision not improved by glasses.
Left eye, 6/12 (2) Hm. +0.75 D.	

Right Eye.—There is a thin transparent veil in the inferior temporal quadrant of the eye. This is attached to the retina on either side of the superior temporal vein by two narrow strands of tissue. The vein at this point is very dark and appears to come forward into the veil but cannot be traced further.

The upper border of the veil is fairly sharply defined with a serrated edge due to small projections of tissue. It lies well forward in the vitreous. In the temporal periphery it ends in a mass of mottled paper-white tissue which appears fenestrated. Terminal branches of the superior temporal artery and the inferior temporal vein can be traced into this tissue, which is very similar to that seen in the vitreous near the ora serrata in the cases of congenital retinal fold described by one of us (I. M.) in the *British Journal of Ophthalmology* in December, 1935.

The veil on its nasal side appears to fuse with the retina as if the latter structure had divided into two layers. It is impossible to define any nasal edge to the veil.

Somewhat external to the region in which the veil and retina fuse, branches of the inferior temporal artery and vein come forward into the veil and end close together in a mass of tissue in which fine vessels (?capillaries) can be seen faintly.

The inferior temporal artery is crossed by a branch of the inferior temporal vein before it enters the veil. There is therefore no doubt that in the first part of its course it runs normally in the retina and only leaves this to enter the veil at the line of attachment to this. The transparency of the veil and the fact that the retina can be seen in position through it shows that the veil is not formed of a double fold of retina but is an additional structure lying in front of this. A branch of the inferior temporal vein slightly peripheral to the branch referred to above comes forward into the veil and ends in a brush of small vessels some of which have terminal knobs. Fig. 1 shows the ophthalmoscopic appearance and Fig. 2 a diagram of the probable arrangement of parts in a vertical antero-posterior section to the temporal side of the disc.

The left eye of the same case showed a fragment of a similar filmy transparent veil floating free in the lower temporal quadrant of the vitreous. The fragment is roughly triangular in shape with

Ophthalmoscopic appearance of the veil and fundus oculi in the right eye of case 1.

FIG. 1.

FIG. 3.

Case 2. A, B and C are vessels invading the veil. Ophthalmoscopic appearance of veil in right eye of case 2.

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FIG. 2

Diagram of antero-posterior section of right eye of case 1 to temporal side of disc.

the base uppermost. The external and internal angles of the triangle are sharp, the inferior obtuse (120° approx.). No connection with the retina was visible nor could any attachment to the lens be seen. On the film are a few fine lines suggestive of obliterated vessels (probably vasa hyaloidea propria). No other abnormality was seen in this eye but the relatively poor visual acuity (6/12 partly) is noteworthy and shows a definite developmental retardation (retinal amblyopia).

Case 2.—Joseph G., aged 20 years, brother of Case 1.

Right, convergent strabismus.

V.A. Right, C.F.: $\bar{C} + 2.0$ D. sph., $+2.0$ cyl., axis 180° , no improvement of vision.

V.A. Left, 6/18. Hm. $+0.75$ D.

Right Eye.—There is a faint white straight streak in the retina starting from the disc about “9 o'clock” sloping upwards and outwards, and fading out in the periphery. About 2D.D. from the disc a branch of a small supernumerary upper temporal artery curves round towards the disc and lies along this streak. At the point of junction of artery and streak (A in Fig. 3) the veil commences as a very narrow strip of tissue projecting only slightly from the retina till it reaches a point close to the disc. There it leaves the retina and comes steeply forward with the artery visible a little deep to its anterior edge but in places obscured by the tissue. The artery contains blood but cannot be traced beyond this point.

The edge of the film which is now far forward in the vitreous,

curves round from in front of the disc towards the temporal side leaving a small section of the inferior temporal quadrant of the disc clearly visible with no intervening veil tissue. This edge appears dark in the sketch (Fig. 3) but this is an optical effect no doubt due to greater thickness of the film at this point. There is no pigment visible anywhere in the veil.

Just after the edge has turned towards the temporal side there are numerous fine pencilled lines visible in the film running from its edge nasally and slightly upwards. These appear to be the remains of obliterated vessels. The veil narrows from this point into a thin bridge connecting the portion in the neighbourhood of the disc with the main body of the film which occupies the periphery of the inferior temporal quadrant of the eye extending a short distance into the upper temporal and the inferior nasal quadrants. This main portion of the film appears to be attached to the globe in the neighbourhood of the ora serrata, and to stretch across the eye in the vitreous not far from the back of the lens. It is completely transparent, the retina being clearly visible beneath it on focusing down. The somewhat opaque appearance of the film in the sketch is due to an attempt on the part of the artist to achieve the difficult task of conveying an impression of the difference of level between the film and the retina. When the film is sharply in focus with a +9 or +10D. lens in the ophthalmoscope the retina can of course be only dimly seen.

Above the bridge, which runs outwards connecting the part of the veil in front of the disc with the main body of the veil, the edge of the latter is sharply defined with several narrow strands of tissue projecting inwards. The lowest of these is the most substantial and appears dark in the sketch. Below the bridge there is a large hole in the veil. This hole is bordered on the nasal side by a vertical bridge of tissue which runs upwards to join the part of the veil which lies in front of the disc. From this vertical bridge several strands of tissue project inwards. Two of these strands join up with the inferior nasal artery. The lower is obviously a branch of the artery and contains blood. The upper appears to be an obliterated branch of the artery (C and B, Fig. 3).

The mass of tissue in the neighbourhood of the disc has no special features other than those already described. It lies in approximately the same plane as the rest of the veil. Its lower border which forms part of the circumference of the hole is sharply defined. Its upper and inner boundaries are more indefinite. The three points at which vessels leave the retina to enter the veil are indicated by arrows in the sketch (A, B and C).

The peculiar fluffy white tissue in the periphery in John G. is not seen in Joseph G. though far out in the upper temporal quadrant is an area in which the choroid presents a peculiar white

stippled appearance. There is no detachment of the retina which can be seen behind the veil over its whole extent.

Left Eye.—Joseph G. In this eye, as in the left eye of his brother, there is a portion of a veil free in the vitreous. There is some retinal amblyopia (6/18). The veil is floating in the lower half of the vitreous, far forward. It is narrow, never more than $1/3$ D.D. in width, but diminishes in places to $1/6$ D.D. Starting from a point below and to the nasal side of the disc it runs horizontally to the temporal side and then bends downwards at almost a right angle, ending below in a sharp point. The horizontal

FIG. 4.

Antero-posterior section of left eye of case 2.

and vertical limbs are of approximately the same length. The edges are well defined. There is no attachment to the retina or lens nor are there any obliterated vascular channels in the veil. The retina is everywhere apparently normal except in the inferior part where there is a shallow detachment, extending to the ora serrata, but without a hole. There is a small limitation of the upper field corresponding to this. The detachment has the smooth appearance of a congenital failure of a portion of the inner layer of the optic cup rather than of a secondary displacement. Fig. 4 shows the relationship of the structures in antero-posterior section.

Case 3. George B., aged 21 years. Right eye, 6/24. Left eye, C. F. The right eye shows hypermetropia (+5.0D.) and a condition of hypermetropic pseudo-neuritis.

The left eye (which has +10.5D. hypermetropia) shows a transparent veil similar to that seen in the right eye of the two previous cases. The veil is attached to the retina along a horizontal line

below the optic disc. Both retinal arteries and veins pass forward on to it and it ends far forward in the inferior periphery in a serrated edge which is fluffy and paper-white and similar to the white tissue at the ora serrata in case 1 and in some of the cases of retinal fold. There are patches of choroidal disturbance in other parts of the fundus. Some are white (as in the upper temporal periphery in case 2) and some pigmented like the scars of old choroiditis. The veil in this case is not quite so transparent as in the others and it is not possible to say that the retina is entirely in position behind it. The general similarity with the other cases, however, warrants the supposition that the condition is the same.

Aetiological considerations. The only acquired pathological condition which is suggested by these cases is retinitis proliferans following haemorrhage into the vitreous. The normal appearance of the retina in the region of the veil, the regular arrangement and the absence of a history of progressive loss of vision and the stationary nature of the condition are against this diagnosis. On the other hand, the presence of choroidal changes in the periphery in two eyes and of a shallow detachment in one might be taken as evidence of past, possibly intra-uterine, inflammation. The occurrence of the veil in both eyes of two brothers is definitely in favour of a developmental origin. It can, therefore, be conceded that the condition is a congenital one. We have then to consider the possible embryological meaning of the appearance. The points calling for special examination are:—

1. The nature and origin of the tissue in the veil.
2. The nature and origin of the vessels in the veil.

In the first place the veil may be a purely abnormal structure lying entirely internal to the inner layer of the optic cup and not containing any retinal structures. In favour of this opinion is its extreme translucency, and the fact that in the left eyes of both John and Joseph G. it is floating free on a plane entirely anterior to the retina which can be seen everywhere continuous behind it. Against this opinion however, is the very striking fact that in the right eyes of John and Joseph G. and in the left eye of George B. vessels running in the veil are quite certainly the continuation forward of vessels which are definitely retinal in the first part of their course. The veil therefore, even if not derived from the inner layer of the optic cup, must have been in continuity with this at some time.

It is possible that the retina has split, a thin lamina from its internal surface forming the veil and therefore naturally containing some retinal vessels. The line of splitting, however, presents some difficulties. Any level deep to the internal limiting

membrane would imply a loss of continuity of conducting neurones and would be represented by a corresponding loss of field, which these cases do not show. If the internal limiting membrane alone had split off, this might account for the appearance and we cannot ignore this possibility, though the veil is somewhat too substantial to be composed of this alone. It seems more probable that the veil is of the nature of a vitreous condensation and that, at its commencement, it was in contact with the inner layer of the optic cup. It may be similar to the vascular vitreous condensations sometimes seen in cases of congenital retinal fold. Fig. 5 shows a case (from the *British Journal of Ophthalmology*, December, 1935), of retinal fold in which a thickened portion of the primary vitreous containing vessels was adherent to the retina and had

FIG. 5.

Ophthalmoscopic appearance of retinal septum in the left eye of a girl of 9 years. A, as seen with a +12 D. sphere. B, as seen without a lens. C, a diagram showing the vertical disposition of the retinal fold attached to the persistent hyaloid. D, a diagram showing the horizontal disposition of the fold. (A=the persistent hyaloid in its glial sheath. B=the scar-like adhesion to the pigment epithelium and choroid. C=the tent-like detachment or fold.)

caused a tent-shaped detachment of this over a narrow area above the disc.

The thickened tissue adherent to the apex of the fold in this case ends far forward near the ora serrata in fenestrated white fluffy tissue similar to that seen in cases 1 and 3 of the present series. It therefore seems justifiable to say that the veil is a vitreous derivate showing some affinity with the abnormal condensations of primary vitreous seen in some cases of congenital retinal fold. A similar tissue is also sometimes seen in cases of persistent hyaloid artery. The tissue is not itself retinal but it may play a part in the production of congenital retinal folds and detachments. It is noteworthy that in case 2 there is a peripheral congenital detachment in the lower part of the retina.*

2. The nature and origin of the vessels in the veil. This is a difficult problem, since the vessels are undoubtedly retinal in one part of their course and vitreous in another. There are three main possibilities. In the first place they may be vasa hyaloidea propria which have become secondarily retinal in a part of their course. Secondly, they may be retinal vessels which have secondarily grown into the vitreous condensation where this is abnormally adherent to the retina. Thirdly they may represent an anastomosis between vasa hyaloidea propria and retinal branches of the arteria centralis retinae. In favour of their being vasa hyaloidea propria which have been laid down on the retina in some part of their course is the fact that in many animals (*e.g.*, rats, mice and some snakes), the definitive vascular supply of the retina is derived from the laying down on it of the membrana vasculosa retinae which is composed of the outer set of the vasa hyaloidea propria. It is therefore quite possible that this might occur as an abnormality over a part of the retina in man. If this were to happen it is probable that no further branches of the arteria centralis retinae would grow into this area as it would already be vascularised at the time when this should be happening.† Indeed cases of partial persistence and attachment of isolated vasa hyaloidea propria to the retina are known in man.

Against this possibility in the present cases is the fact that in case 1 a retinal vein crosses superficial to the artery while it lies on the retina before entering the veil. This would not be likely if the vessel were a vitreous one laid down on the retina as then it should be morphologically superficial to the internal limiting membrane while the veins, developed later, would be deep to this.

* Embryologically speaking, the term "detachment" is wrong. "Failure of coaptation" is more correct.

† The vasa hyaloidea propria are at the height of their development by the 48 mm. stage, the branches of the central artery only appear at 100 mm.

FIG. 6.

Possible modes of production of vascular veils. A. One of the vasa hyaloidea propria in the lower part of the eye becoming retinal in the first part of its course. B. A branch of the arteria centralis retinae running into the vitreous veil. C. An anastomosis between a branch of the central artery and one of the vasa hyaloidea propria. H. Hyaloid. V.h.p. Vasa hyaloidea propria. R. Retinal artery. B. Bulb of hyaloid.

On the whole this explanation does not correspond with the appearances.

There is more to be said in favour of the second possibility that the retinal vessels have grown into the veil secondarily. If a condensation in the primary vitreous had remained adherent to

the retina at one point without exerting sufficient traction to produce a congenital retinal fold by hindering coaptation of the two layers of the optic cup, then it is likely that it would become vascularised by a retinal vessel. It is well known that vessels of new formation can bud out of retinal vessels into the vitreous (*e.g.*, in retinitis proliferans, in Lindau's disease and in organising vitreous haemorrhages) in adult life, and there is no inherent impossibility that, at the 100mm. stage when the branches of the bulb of the hyaloid on the disc are invading the retina they might not also invade abnormally placed strands of primary vitreous if this were closely adherent to and in continuity with the retina. It is possible that since the adhesion might antedate the formation of the membrana limitans interna the proper development of this might be interfered with in the region of the adhesion and so the retina would not be sharply demarcated here. (The absence of Descemet's membrane in the region of a congenital anterior synechia is a similar phenomenon.)

This is consistent with the facts that the arrangement of the retinal vessels seems perfectly normal elsewhere and that the extent of invasion of the veil by retinal vessels is very different in the various eyes.

The third possibility that there is, along the line of attachment of the veil, an anastomosis between retinal vessels and vitreous vessels, is not likely, chiefly on account of the time factor. The vasa hyaloidea propria are all atrophied before the branches of the arteria centralis retinae have extended from the disc into the retina. It would only be in exceptional cases that they would persist long enough to form an anastomosis.

On the whole therefore the second possibility is the most likely. Fig. 6 shows some of the embryological possibilities.

The initial cause and exact mechanism is impossible to state but the cases seem to throw some light on possible anomalies of the primary vitreous and their effects on the retina.

REFERENCE

- IDA MANN.—Congenital retinal fold. *Brit. Jl. of Ophthalm.*, December, 1935, pp. 641-658.