Posterior hyperplastic primary vitreous

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SUMMARY A fundus picture of posterior hyperplastic primary vitreous based on a study of 14 cases is described. Its definition, in spite of pleomorphic manifestations, could be useful for its differentiation from other conditions affecting children's eyes and presumed to be caused by parasitic, inflammatory, fibroplastic, or genetically transmitted processes.

The basic role of persistent hyperplastic primary vitreous in the aetiology of tunica vasculosa lentis and its complications has been well established for nearly a century. Ample histological material was provided as children's eyes were being enucleated because of an erroneous diagnosis of retinoblastoma. In 1935 Ancona (1935), Mann (1935), and Weve

(1935) reported on congenital falciform retinal folds (ablatio falciformis retinae, congenital retinal septum), and Mann (1935) advanced a concept—based on embryological studies—of the basic aetiological role of persistent primary vitreous in this condition too. Mixed cases were found in which both anterior and posterior segments seemed to be involved, and the reports were the subject of scholarly review by Reese (1955) in his Jacksonian lecture. The responsibility of hyperplastic primary vitreous for the causation of certain retinal holes

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Fig. 1 Case 1. a. The right eye of a 7-year-old girl seen 4 years ago. Visual acuity 6/18. Glial tissue partly covers the optic disc and extends as a strand up and nasally to end in a white, hazy knob at the surface of the retina. The macula (not in the picture) showed cystic degeneration and tension striae towards the disc. b. The same eye 4 years later. Visual acuity 6/36. Marked increase in elevation of glia at the disc. A thick fold now stretches from the disc downwards carrying retinal vessels. Nasally several fine linear strands are seen (? primary vitreous), as well as deep pigmentary disturbance.

Fig. 2 Case 2. The left eye of an 8-year-old boy. Visual acuity 6/36. The optic disc is shapeless; its true outline is seen only at the 12 o'clock meridian. A white saucer-like plaque extends from there down and temporally. The blood vessels form hairpin bends and arcades. Deep pigment dystrophy and abnormal deep glial tissue is seen

Fig. 3 Case 3. The left eye of a 6-year-old boy. Visual acuity counting fingers. A ragged white deposit is nearly totally masking the optic disc. Two folds extend from it downwards and upwards towards the periphery. Temporally the retina is pulled forwards; a similar but lesser pull is seen at the 10 o'clock meridian. The vessels are tortuous and form hairpin bends

Fig. 4 Case 4. The right eye of a 5-year-old boy. Visual acuity perception of light. A dense, ovoid, saucer-shaped plaque covers the macula. The blood vessels are distorted and stretched. They form hairpin bends. Tension folds radiate towards the inferotemporal periphery. The optic disc is partly covered by glial tissue, which merges temporally with the macular plaque. It shows no physiological cupping and is fresh coloured. Along the stretched blood vessels are glistening white tissue strands (? primary vitreous)

and retinal detachments was suggested by Hagedorn and Sieger (1956) and Pruett and Schepens (1970).

The presence of abnormalities in the posterior segment of the eye, even without the anterior segment being affected, was established by Manschot

(1958) and Wolter and Flaherty (1959) as a result of careful histological studies. Pruett (1975), reviewing his clinical material from 30 patients (33 eyes), stressed the variety of fundus lesions. The following clinical features were found in his series:

Fig. 5 Case 5. The right eye of a 6-year-old girl. Visual acuity counting fingers. A fist-like, dense white band arises from the optic disc and extends over the macula, with starfish folds radiating temporally. The blood vessels are tortuous and form arcades. Nasally there is a concentric halo of streaky glistening deposits (? primary vitreous)

Fig. 6 Case 6. The left eye of a 10-year-old girl. Visual acuity perception of light. The optic disc is flesh coloured, and shows no physiological cupping. A veil-like band of white-greyish tissue covers its temporal part and extends and runs over the macula, where it ends. The macula shows a pigment clump surrounded by a depigmented halo. The blood vessels form scalloped arcades and hairpin bends. Inferiorly the retina shows a metallic reflex (? primary vitreous)

Fig. 7 Case 7. The right eye of a 5-year-old girl. Visual acuity 6/60. The optic disc is of flesh colour and shows no physiological cupping. From it a fold of elevated retina reaches the macula, which is masked by a ragged plaque of white tissue, with spiky extensions from its temporal edge. The blood vessels show tortuosity superiorly and are stretched inferiorly. A fine glistening pattern is seen in the upper periphery of the picture (? primary vitreous), and a patch of deep pigment disturbance below the disc

Fig. 8 Case 8. The right eye of an 18-year-old girl. Visual acuity 6/60. From a distorted, flesh coloured, and ill-defined disc strands of white veil-like tissue reach the macula, where they terminate as a furry, dense, white knob. From there similar strands and veils extend upwards. They are preretinal but posterior to the vitreous and not elevated, and they distort and pull the retinal vessels with them. Fluorescein angiography showed a glow from the optic disc

retinal detachment, 15 eyes; macular pigmentation, distortion, and involvement by a fold, 23 eyes; vitreous haemorrhage, 1 eye; glaucoma, 1 eye; cataract, 2 eyes. As these children were referred to the Boston Retina Unit, internationally reputed for its retinal detachment work, the referrals were likely to have been selective.

The present report is based on 14 cases seen during the past 8 years in the diagnostic angiography unit, which by its nature did not attract the referral of patients affected by retinal detachment or anterior segment lesions. The clinical picture which emerged from our study is presented to help in the diagnosis of persistent posterior hyperplastic primary vitreous (PHPV), which may be difficult in view of pleomorphic and often bizarre manifestations of its presence.

Patients and methods

Of the 14 patients 8 were male and 6 were female. Their ages when they were first seen were between 5 and 25 years (11 patients 5–10 years, 3 patients 15–25 years). In 13 cases only 1 eye was affected; in 1 case the other eye was affected to a lesser degree (case 14).

The reasons for primary referral to an ophthalmologist were poor unocular vision or a squint. The levels of visual acuity (15 eyes) were as follows:

Fig. 9 Case 9. The right eye of a 10-year-old boy. Visual acuity hand movements. **a.** The optic disc consists of a white, wide rim seen only on the nasal side and a small, central, flesh coloured core. It is continuous temporally with a similar rim of slightly larger diameter to form an oval saucer which encompasses the macula and disc. From the centre of this saucer arises a grey-whitish cone of glia. The periphery shows widespread deep pigmentary disturbance. **b.** The far periphery shows glistening linear deposits (? primary vitreous)

Fig. 10 Case 10. The left fundus of a 6-year-old boy. Visual acuity perception of light. The disc-macula complex is difficult to localise with precision because of gross distortion of all central retina by flat preretinal bands. There are dense, white deposits, the lower one probably at the macula. The blood vessels form hairpin bends; some are tortuous, and some are stretched into narrow, straight course. The periphery shows widespread deep pigmentary disturbance. The other eye has 6/6 vision; its fundus showed marked tortuosity of vessels only

Fig. 11 Case 11. The left eye of an 8-year-old boy seen 7 years ago. Visual acuity counting fingers. a. The outline of the disc is lost under a thick preretinal membrane and glial deposits. The disc is flesh coloured. The central vessels are tortuous and form hairpin bends. A thin fold-like band runs from the disc up and nasally. From a dense tulip-like deposit on the temporal side of the disc another band runs out and temporally. This temporal band thins out and spreads to end below the raphe

Fig. 12 Case 12. The left eye of a 10-year-old girl. Visual acuity counting fingers. a. The optic disc is distorted, shows no physiological cupping, is flesh coloured, and has a hazy outline. The blood vessels show marked tortuosity, uneven calibre, and crowding. The macula is covered by a membrane. b. Fluorescein angiography shows that in spite of gross tortuosity there are no leakages from the blood vessels. c. The optic disc shows marked late stain typical for glial and fibrous tissue

Fig. 13 Case 13. The right eye of a 25-year-old man. Visual acuity 6/60. The optic disc shows flesh colour, no physiological cupping, and its outline is defined only on the nasal side. Superiorly the blood vessels are tortuous, distorted, and unevenly covered by a preretinal membrane. This membrane extends over the macula. Fluorescein angiography showed late stain of the optic disc

6/9, 1; 6/18–6/60, 6; counting fingers to perception of light, 8. All fundi were photographed, but fluorescein angiography could not be performed efficiently in most patients because of their age. In fact only 6 patients had this test. All were investigated for parasitic, inflammatory, and autoimmune disease, with negative results. In no case did we find any of the features characteristic of anterior segment involvement—for example, microcornea, angle abnormalities, cataract, retrolental membrane, cloudy vitreous, glaucoma. No eye was inflamed, irritable, or photophobic.

Results

The legends of Figs. 1–14 describe the pathological findings in individual cases. The incidence of salient features is summarised in Table 1. Many individual features were present simultaneously in affected eyes. However, the dominating features for the series can be established.

Thirteen cases were unilateral (93%). The optic disc was severely affected in all of them by distortion, loss of outline, absence of physiological cupping, and fleshy disc colour. 53% of the eyes showed radiating folds and bands, and 47% showed gross involvement of the disc-macula complex by heavy

Fig. 14 Case 14. The left eye of a 15-year-old boy. Visual acuity 6/18. a. The blood vessels are markedly tortuous. At the 11 o'clock meridian several small vessels are stretched in the horizontal direction, ladder-like. The superior temporal vein is covered by a grey membrane just above the disc. The macula is covered by a grey thin membrane, and above it the blood vessels are markedly scalloped, forming arcades. The optic disc is poorly defined, fleshy, and shows no physiological cupping. The other eye showed nearly symmetrical changes, but the macula was free of membrane, with vision 6/9. b. Fluorescein angiography shows no leakages from blood vessels but late glow of the optic disc

Table 1 Incidence of individual features (15 eyes)

<i>Disc</i>		
Distortion, loss of outline	15	100%
Absence of physiological cupping	15	100%
Fleshy colour	15	100%
Glial proliferation	11	73%
Radiating folds and bands	8	53%
Gross disc-macula deposits	7	47%
Late fluorescence	all 7 tested	
<i>Macula</i>		
Preretinal membrane	10	67%
Glial deposits	7	47%
Cystic degeneration	4	27%
Pigment dystrophy	4	27%
Displacement	2	13%
<i>Vessels</i>		
Distortion	13	89%
Tortuosity	12	80%
Hairpin bends	8	53%
<i>Periphery</i>		
Deep pigment disturbance	9	60%
Glistening deposits	6	40%

deposits of glial tissue. All 6 cases which had fluorescein angiography showed marked, late fluorescence of the disc, case 14 in both eyes.

The maculae were affected secondarily in all cases by premacular membrane, glial deposits, pigment dystrophy, or cystic degeneration. The retinal blood vessels were distorted, tortuous, and formed hairpin bends in all cases. It is interesting that in 6 cases (40%) peripheral glistening deposits were seen,

some similar to those reported by Pruett (1975). Deep pigment disturbance outside the macular area was seen in 60% of cases.

Discussion

These observations indicate a pleomorphic fundus syndrome caused by the persistence of primary hyperplastic vitreous in the posterior segment of the eye. The occurrence of retinal detachment, as reported previously by other workers, has to be incorporated to make the syndrome complete. The differential diagnosis is between retrolental fibroplasia, parasitic infections, inflammatory conditions, and congenital vascular abnormalities of the retina.

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