Original Article

A clinicopathological study of persistent fetal vasculature

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Purpose: To study the clinicopathological findings of Persistent Fetal Vasculature (PFV) in patients with congenital cataract and PFV. **Methods:** Six eyes with anterior or combined PFV with cataract underwent phacoaspiration with primary posterior capsulotomy with anterior vitrectomy with intraocular lens implantation followed by histopathological evaluation of the PFV stalk and membrane. **Results:** Four and two patients had combined and anterior PFV respectively. There was no postoperative hyphema, vitreous haemorrhage, glaucoma or retinal detachment in six months. Haematoxylin and eosin staining showed inflammatory cells predominantly with extramedullary hematopoeisis and vascularisation. **Conclusion:** We recommend IOL implantation in PFV, with early and aggressive amblyopia therapy.

Key words: Congenital cataract, histopathology of persistent fetal vasculature, persistent fetal vasculature, persistent hyperplastic primary vitreous

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Persistent fetal vasculature (PFV) [persistent hyperplastic primary vitreous (PHPV)] is a rare ocular disorder, first described by Reese.[1] It is usually unilateral, inherited sporadically without any systemic association.[2]

PFV is characterized by cataract, microphthalmos, vascularized retrolental tissue, and elongated ciliary processes [Fig. 1].[1] It can be anterior, posterior, or combined (the most common type),[1] diagnosed clinically and ultrasonographically, showing retrolental mass from optic disc to posterior capsule without calcification.[3] Phacoaspiration with or without intraocular lens (IOL) implantation is the primary treatment with intraoperative bleeding due to vascularization a major challenge.[4] Retinal detachment, glaucoma, and amblyopia cause poor prognosis.[5]

In this series, we aim to study the characteristics of PFV stalk and membrane on histopathology, since it might help in diagnosis and prognostication.

Methods

This was a prospective case series done at a tertiary hospital in India from October 2015 to June 2017, after Institutional Review Board approval. The study adhered to the tenets of Declaration of Helsinki. We included six eyes with unilateral cataract and anterior or combined PFV, without trauma, retinal detachment, or glaucoma. Detailed ocular examination was done under general anesthesia, with B-scan ultrasound. IOL power was calculated by SRK-T formula/Dahan guidelines. All patients underwent phacoaspiration, primary posterior capsulotomy, and anterior vitrectomy with or without IOL implantation under general anesthesia. After making ports, anterior chamber was formed by 1.4% sodium hyaluronate. Anterior capsulorhexis was followed by hydrodissection, bimanual irrigation-aspiration, and capsular polishing. Posterior capsulorhexis and anterior vitrectomy were done with vitrectomy cutter. The vascularized stump and plaque were cauterized, resected in toto by scissors, and sent for histopathological examination in 10% neutral buffered formalin. A foldable IOL was implanted. At 2 weeks postoperatively, retinoscopy was prescribed with patching of the dominant eye for amblyopia management.

Results

All our patients were males with age <3 years (minimum 2 weeks), with white reflex as chief complaint. One patient had contralateral anophthalmic socket. None of our patients had systemic or family history. Preoperatively, two patients had central, steady, maintained (CSM) fixation, according to CSM method. Two patients had uncentral, unsteady, maintained fixation, while two had uncentral, steady, maintained fixation. On ultrasonography, four and two patients had combined and anterior PFV, respectively. None of our patients had microphthalmos. The lowest and highest corneal diameters recorded were 9 and 11.5 mm, respectively (mean 10.08 mm). All patients had vascularized plaque on posterior capsule. IOL was implanted in bag and in sulcus in four and one

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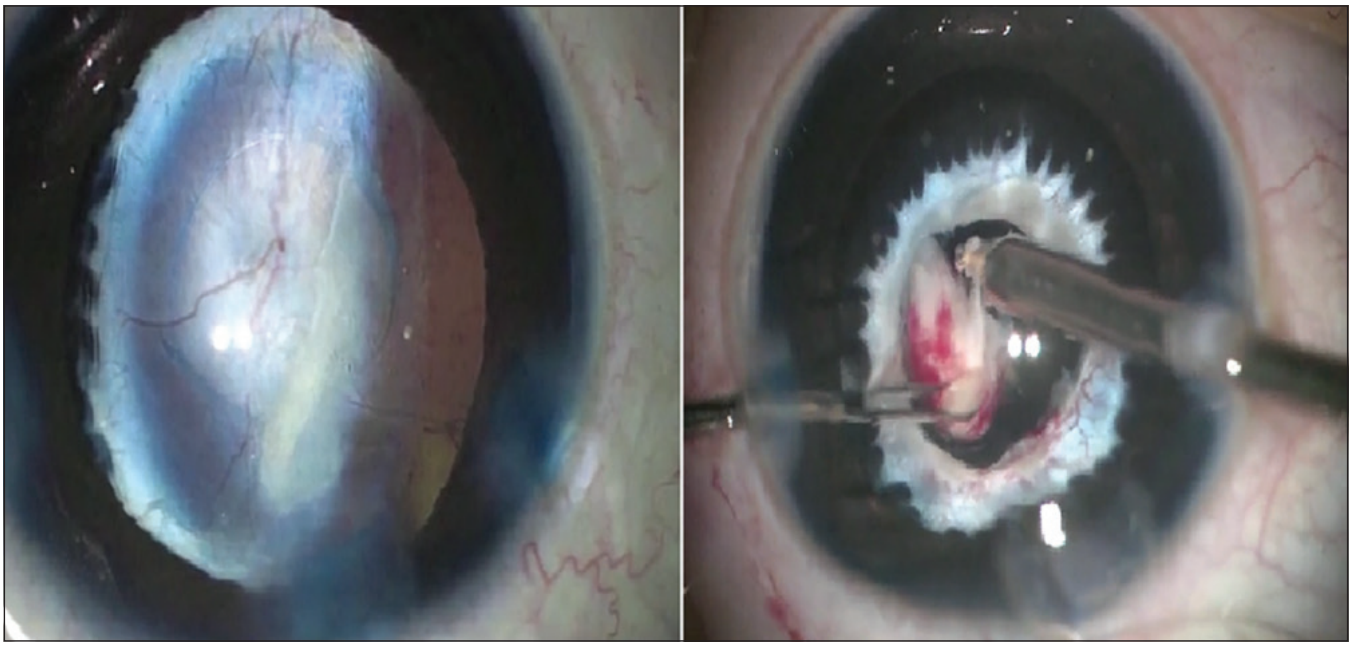

Figure 1: Preoperative photograph (*left*) with a white, dense vascularized membrane along with enlarged ciliary processes temporally and intraoperative photograph (*right*) showing resection of the vascularized membrane after cauterization

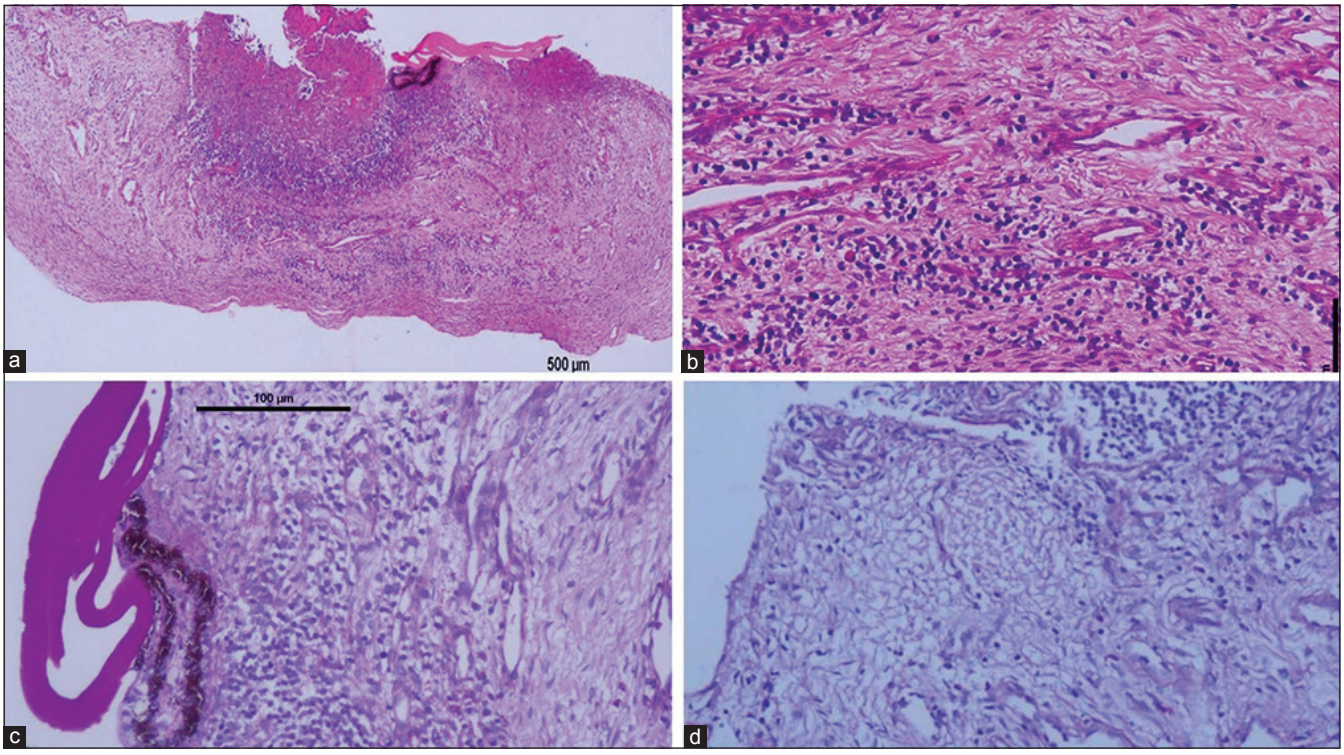

Figure 2: (a) Low-power photomicrograph showing full-thickness biopsy composed of highly vascularized mesenchymal cells and collections of inflammatory cells (hematoxylin–eosin, ×50). (b) High-power photomicrograph to bring out the loose mesenchymal tissue and thin-walled blood vessels and the type of the inflammatory cells, dominantly mononuclear cells along with eosinophil precursors (hematoxylin–eosin, ×400). (c) High-power photomicrograph showing fibrinous exudate, pigmented cells (part of ciliary body), band of amorphous eosinophilic material (fragment of lens), and moderately heavy infiltration by inflammatory cells dominantly mononuclear cells. The vascularized component is also seen better (hematoxylin–eosin, ×400). (d) High-power photomicrograph showing the band of amorphous eosinophilic material (fragment of lens) taking up strong periodic acid–Schiff positivity and the linearly arranged pigmented cells (periodic acid–Schiff, ×400)

patients, respectively. One patient was left aphakic due to microcornea (9 mm). Three-piece and single-piece hydrophobic acrylic IOL were implanted in one and four patients, respectively. At 6 months, five patients had CSM vision and

one patient had central, unsteady, unmaintained fixation. All patients had clear visual axis in the early postoperative period, obscured by posterior capsular opacification (PCO) later in two cases, requiring membranectomy at 2 weeks and 1 month. There was no hyphema, vitreous hemorrhage, glaucoma, or retinal detachment in any patient in 6 months.

Hematoxylin and eosin staining was done to evaluate the microscopic sections of the vascularized stalk. Light microscopy showed inflammatory cells predominantly in low magnification. Mononuclear cells with eosinophil precursors such as promyelocytes, myelocytes, and metamyelocytes, with vascularization, loose mesenchymal tissue and multiple thin-walled blood vessels were seen in medium and high magnifications. Mesodermal hyperplasia with smooth muscle actin was present in vascular channels' walls, seen as loose mesenchymal fibrovascular connective tissue. Congested vascular channels, diffusely infiltrated with erythroid and eosinophilic precursors, suggesting extramedullary hematopoiesis were seen.

Discussion

PFV is caused by failure of primary vitreous and persistent hyaloid vasculature regression,[1] seen as a retrolental stalk. Haddad *et al*. report leucocoria, microphthalmos, and cataract as the most common presentation, seen in our study also.[6] All patients had unilateral disease. Incidence of bilateral PFV is < 10%.[7] All our patients were males and none had microphthalmos, not consistent with other studies.[8] PFV has never been reported and confirmed on histopathology in age as less as 2 weeks, as seen in our study. A-scan and B-scan ultrasonography should be done for diagnosis and axial length for microphthalmos and IOL power calculation.

In our series, combined PFV was more common. Combined PFV has poorer visual prognosis than anterior PFV.[5] We had taken vitreous stalk and vascularized membrane in combined and anterior PFV, respectively, for histopathology. Histopathology confirmed the diagnosis. PFV is known for intraoperative bleeding, postoperative hyphema, and vitreous hemorrhage, which were avoided in our study due to cauterization.[4] We corroborate these complications to extensive vascularization, thin blood vessels, erythroid precursors, and extramedullary hematopoiesis, as seen in our study [Fig. 2a–d] and other literatures.[6,9] Inflammatory cells might be cause for increased PCO. Amount of vascularization and fibrous component can be compared. More vascularization will increase bleeding, hyphema, and vitreous hemorrhage and fibrous component will increase retinal traction and detachment. So, histopathology might prognosticate a case and individualize the follow-up. Enlarged, pigmented, and serrated ciliary processes, pathognomonic sign of PFV,[10] were seen in all our patients clinically and on histopathology. Anisometropic amblyopia is the most common cause for poor vision in these patients, but responds to amblyopia therapy.[4]

Conclusion

We recommend IOL implantation in PFV, with early and aggressive amblyopia therapy. Limitations of our study were short follow-up of 6 months and lack of immunochemistry.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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