Unilateral Pediatric Cataract Secondary to Atypical Anterior Persistent Fetal Vasculature: A Case Report

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ABSTRACT

Persistent fetal vasculature (PFV) is a congenital disorder resulting from the failure of regression of the vasculature of the primary vitreous and hyaloid, leading to a wide clinical spectrum generally classified into anterior, posterior, and mixed types. The anterior type is associated with cataract, posterior capsular opacity, fibrovascular membrane behind the lens, and ciliary body elongation, while the posterior type is related to fibrovascular stalks connecting the vitreous vascular membrane and the optic disc. Recognizing the clinical features and spectrum of PFV, particularly that of the anterior type and including its possible atypical presentations, is essential in pediatric cataract surgeries for surgical planning and postoperative management. This report aims to discuss the diagnosis and pathogenesis of anterior PFV and describes a case presenting with an intralenticular membrane and the absence of ciliary process elongation — two atypical features that have never been reported to coexist in a single case.

A four-year-old male presented with a unilateral, sporadic cataract of the left eye, first noted at three years of age, associated with esotropia. Initial clinical examination and B-scan ultrasonography did not support a diagnosis of PFV preoperatively. However, intraoperative findings revealed an anterior PFV with atypical features. These included an intralenticular location of the anterior PFV and the absence of ciliary process elongation or anterior segment abnormalities on postoperative ultrasound biomicroscopy. Despite being atypical, these findings can be explained by proliferation from the posterior capsule to the lens, leading to the intralenticular location, and continued regression, resulting in the absence of anterior segment abnormalities. This is the first reported case presenting with both of these atypical features.

This case illustrates that anterior PFV can manifest with rare, atypical features, specifically an intralenticular membrane and the absence of ciliary process elongation, likely due to continued vascular regression. Recognizing this variable spectrum is essential in preoperative planning. Routine evaluation for PFV in unilateral, sporadic pediatric cataracts using ultrasound biomicroscopy is recommended to detect such anomalies. Identifying these features preoperatively is crucial for anticipating intraoperative complications and optimizing surgical outcomes

Keywords: persistent fetal vasculature, pediatric cataract, ocular ultrasonography, anterior segment abnormalities, congenital eye anomalies, case report

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INTRODUCTION

Persistent fetal vasculature (PFV), previously known as persistent hyperplastic primary vitreous, is a congenital ocular disorder resulting from the failure of regression of the vasculature of the primary vitreous and hyaloid.[1-3] Most cases of PFV are sporadic, unilateral, and not associated with systemic conditions.[4] Due to its mechanism of development, the resulting remnants create a wide spectrum of pathology,

leading to a constellation of clinical features.[5,6] This variation may even include cases where the hyaloid vasculature undergoes regression after the pathology has already developed.[6]

PFV is broadly classified into anterior, posterior, and mixed forms. The anterior type (25% of cases) is defined by cataract, retrolental fibrovascular membrane, and ciliary body elongation.[2,7] The posterior type (12%) involves fibrovascular stalks connecting the optic disc to the vitreous membrane, often with retinal abnormalities.[2] The mixed type, exhibiting features of both, comprises the majority (62%).[2]

PFV is frequently associated with cataracts, presenting surgical challenges due to increased risks of retinal detachment, hyphema, and glaucoma.[1] Detecting PFV preoperatively is critical for surgical planning; however, media opacification often necessitates ancillary diagnostics like B-scan ultrasonography, CT, or MRI to confirm the diagnosis.[8]

Management remains controversial due to the spectrum of pathology, but surgery is generally recommended to prevent complications such as amblyopia and phthisis.[9,10] While outcomes for anterior PFV are generally favorable, the risk of intraoperative bleeding from vascular remnants remains a significant concern.[1,4,10,11]

This case report describes a four-year-old male initially diagnosed preoperatively with developmental cataract based on clinical and ancillary examinations. Surgical management involved lens extraction with intraocular lens (IOL) implantation, during which findings suggested a diagnosis of anterior PFV.

Postoperative examinations ruled out other pathologies but confirmed two distinct, atypical features: an intralenticular location of the fibrovascular membrane and the complete absence of ciliary process elongation. To the best of our knowledge, these two atypical features have never been reported to coexist in a single case. This report details the diagnostic challenges, surgical techniques, and outcomes of this unique presentation, highlighting the importance of recognizing such variations in pediatric cataract surgery.

CASE PRESENTATION

A four-year-old male accompanied by his mother consulted due to a one-year history of worsening leukocoria in the left eye. The history of illness indicated an onset of a small whitish lens opacity in the left eye one year prior, which subsequently enlarged and worsened, accompanied by increased sensitivity to bright light. No prior medical or surgical interventions were administered for the condition. No associated symptoms such as nystagmus, eye pain, eye redness, tearing, or blepharospasms were observed.

The review of ancillary history revealed an unremarkable pre- and peri-natal course, with no maternal insults, feto-maternal complications, or hospitalizations noted. Past medical history was unremarkable, with no known allergies or history of ocular trauma. Family history was negative for congenital cataracts, blindness, or other hereditary ocular and systemic diseases. The patient lives with his parents and demonstrates age-appropriate psychosocial development.

The patient's best corrected visual acuity (BCVA) was 20/20 in the right eye, but only hand movement with good light projection was noted for the left eye. Dry objective refraction measured +1.50 diopters (D) for the right eye, with just a poor streak noted for the left eye. No relative afferent pupil defect (RAPD) nor pupillary reaction abnormalities were observed, despite the lens opacity. However, a fifteen-degree esotropia by Hirschberg and a 25 prism diopters (PD) esotropia by Modified Krimsky were measured, both with the right eye fixating.

Slit lamp examination of the left eye revealed a cataractous lens with diffuse multiple whitish cortical and nuclear opacities within, with no visible fibrovascular structures within or behind the lens, as illustrated in Figures 1A and

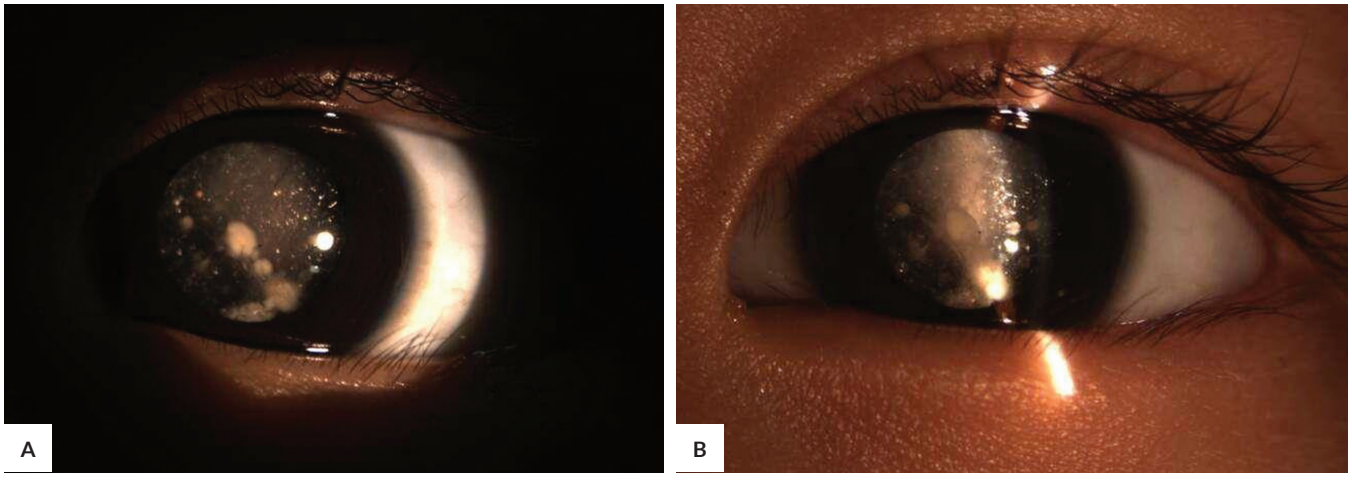

Figure 1. (A and B) Slit lamp photos of the left eye revealing a cataractous lens with no obvious fibrovascular membrane within or behind the lens.

1B. There were no associated corneal opacities, uveitic signs, dilated iris vessels, ectropion uveae, or shallowing of chambers noted. Slit lamp findings for the right eye were unremarkable. Tonometry measurements indicated no hypotony, recording 15 mmHg for the right and 16 mmHg for the left. Fundus examination of the left eye was not possible due to media opacity, but it was unremarkable for the right eye, with no fibrovascular stalk observed.

From the history and clinical findings, an assessment of developmental cataract with associated sensory esotropia was made. Although all necessary diagnostic modalities were available within the institution, the patient faced financial barriers to access; these were resolved through the assistance of Medical Social Services. The B-mode ultrasound examination revealed an intact lens with a low-amplitude thin curvilinear band echo with good aftermovement, implying a posterior vitreous detachment with few vitreous condensations, as shown in Figure 2. No amplitude echoes behind the lens extending to the optic nerve were noted. The estimated axial length of the left eye on the ultrasound was 23.4 mm, supporting the absence of microphthalmia. Given the low consideration of posterior pole and other associated pathologies, surgery was indicated to clear the visual axis, accompanied by a contemplated post-operative amblyopia therapy.

During surgery, routine irrigation and aspiration of the cataractous lens, including its cortical material, was performed. However, completion of the aspiration revealed a posterior fibrous membrane closely adherent to the posterior capsule located superotemporally, as shown in Figures 3A and 3B. Despite attempts at aspiration and manual dissection, the membrane remained tightly adherent to the posterior capsule.

A combination of hydrodissection, aspiration, and manual dissection was utilized, revealing vascularities deep within the membrane with subsequent bleeding on further manipulation, demonstrated in Figures 4A to D. With the clear view established from the aspiration of the cataractous

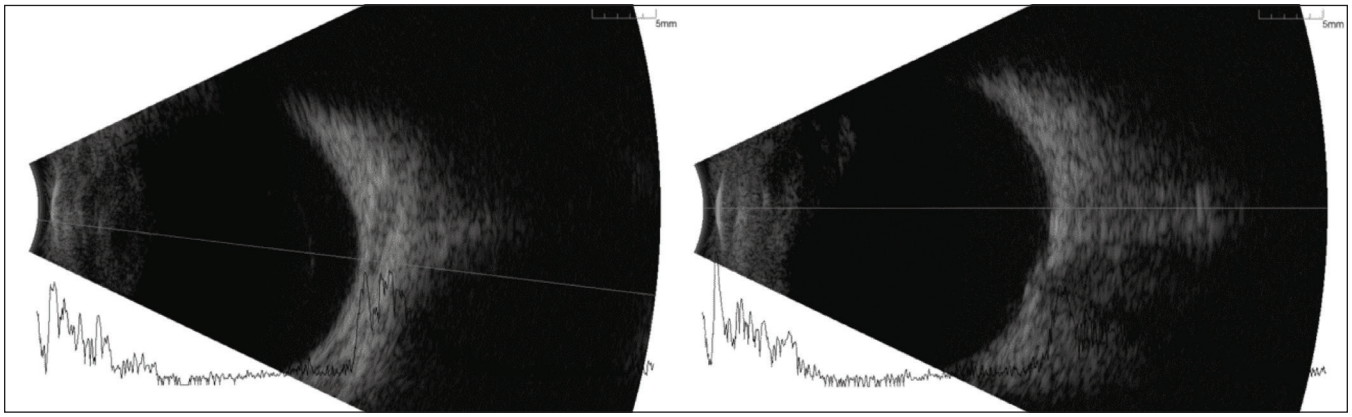

Figure 2. B-mode ocular ultrasound of the left eye showing an intact lens with posterior vitreous detachment and vitreous condensations.

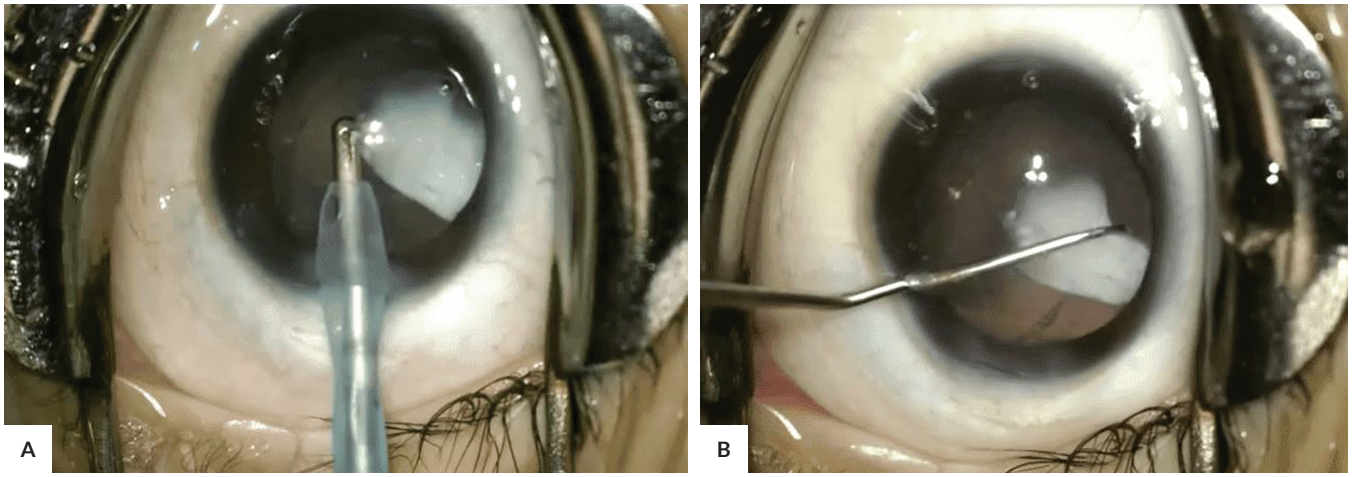

Figure 3. (A and B) Upon aspiration of the cortical material, a fibrous membrane was seen tightly adherent to the posterior capsule, resistant to both attempted aspiration and manual dissection.

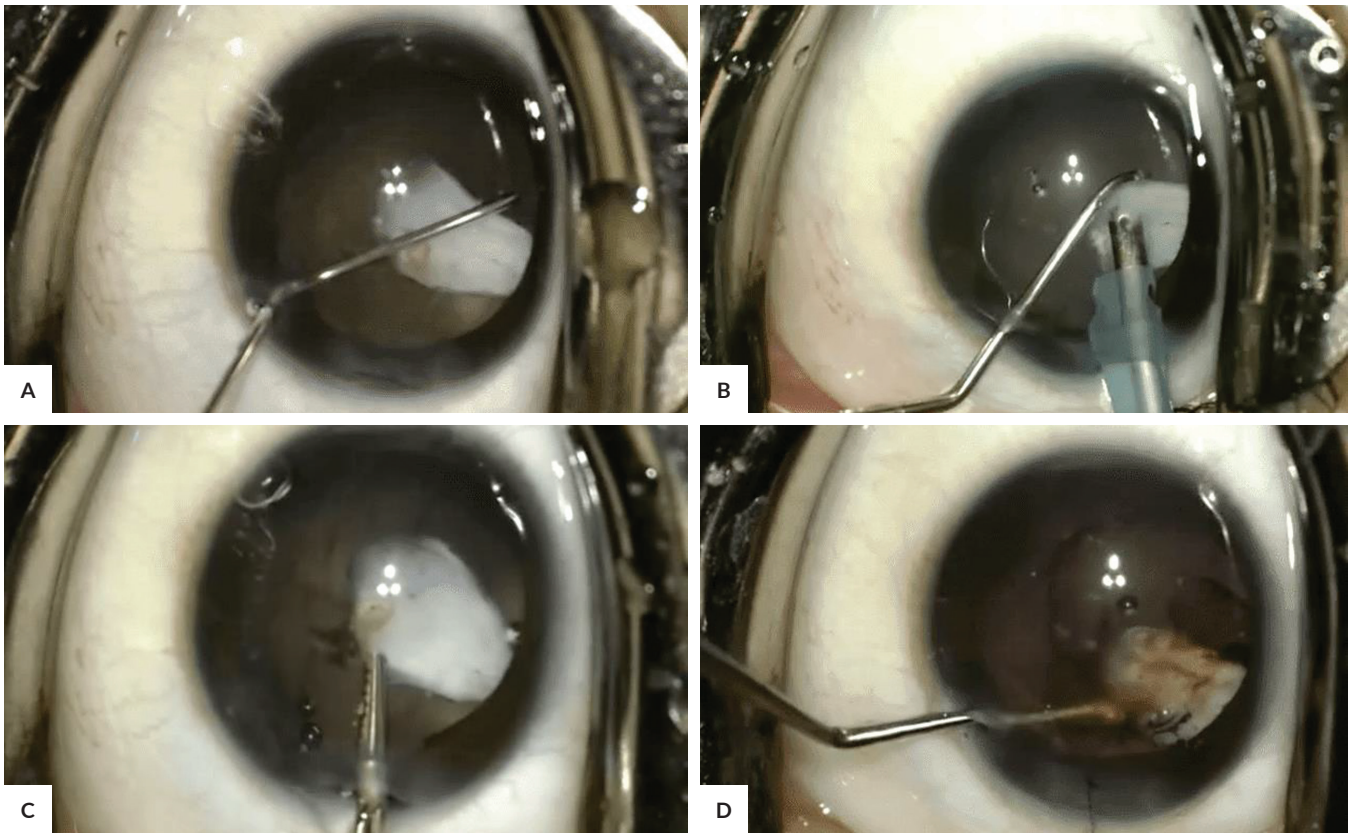

Figure 4. (A to D) A combination of hydrodissection, manual dissection, and aspiration of the fibrous membrane was attempted, with the exposure of deep vascularities and subsequent hemorrhage on manipulation.

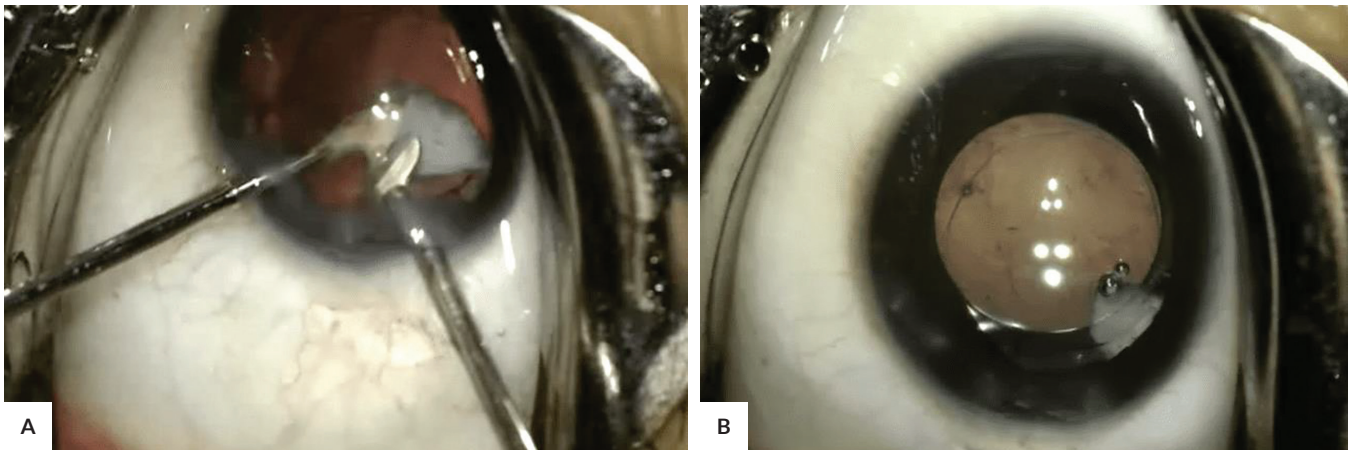

Figure 5. (A and B) Clearance of the central visual axis was done by partial excision of the fibrovascular membrane and its subsequent tamponade via intraocular lens implantation.

lens, an intraoperative fundus examination was conducted which revealed no connections of the fibrovascular membrane to the optic disc or to the rest of the retina, including its periphery. No apparent connections to the ciliary processes were observed intraoperatively either.

Due to the subsequent bleeding on further manipulation and an unknown source of vasculature, the surgical plan was shifted from complete peeling of the fibrovascular membrane to partial excision just enough to clear the central visual axis. This was followed by tamponade via IOL implantation, as exhibited in Figures 5A and B.

Immediate postoperative findings for the left eye included an improvement in vision to at least finger play at 1 foot, a dry objective refraction of +2.50 D, an in-place

IOL with a clear central visual axis, minimal activity of trace cells, and normotonic pressure. A fundus examination revealed a good red-orange reflex with no vitreous hemorrhage or fibrovascular stalk seen, as shown in the fundus photo in Figure 6.

Ultrasound biomicroscopy (UBM) performed three months post-operatively revealed a deep anterior chamber, flat iris configuration, correctly positioned ciliary bodies and ciliary processes, with no iridotrabecular contact, process elongation, or possible sources of vascularization, as recorded in the scans shown in Figure 7.

Four months postoperatively, with good compliance to amblyopia therapy, clinical findings for the left eye included an improved BCVA of 20/50, with no pupillary abnormalities or RAPD. Slit-lamp findings at this time included a formed chamber, an in-place IOL, with a whitish opacity located at the one o'clock position with non-obscuration of the central visual axis, and absence of anterior chamber activity. Given the successful anatomical restoration and good initial response to therapy, the visual prognosis is favorable, though guarded by the risk of amblyopia and secondary capsular opacification.

The summary of the clinical course and management is detailed in Table 1.

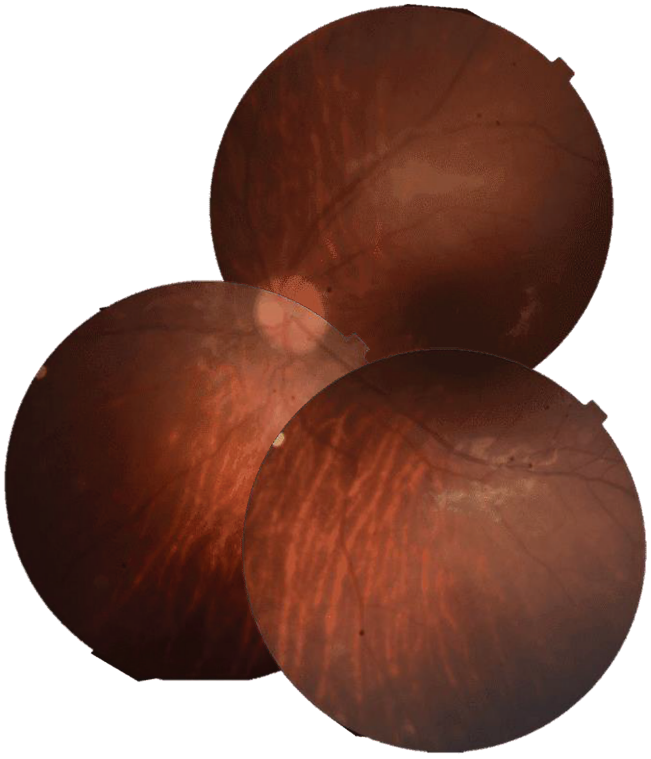

Figure 6. Reconstructed fundus photo of the left eye showed absence of a fibrovascular stalk emanating from the optic disc and absence of postoperative hemorrhage. Peripheral retina is not shown but also did not demonstrate any vascular connection with the posterior capsule.

Table 1. Timeline of the Case

Time	Event
1 Year Prior to Consult (Age 3)	Parents noted onset of small whitish lens opacity in the left eye.
Interim	Progressive enlargement of opacity accompanied by photophobia.
Consultation (Age 4)	Presentation with leukocoria. Diagnosis of developmental cataract. B-scan showed intact lens with no posterior anomalies.
Surgery (Day 0)	Lens extraction performed. Intraoperative discovery of fibrovascular membrane adherent to posterior capsule. Partial excision and IOL implantation performed.
Immediate Post-operative	Vision improved to finger play. Fundoscopy confirmed absence of posterior connections.
3 Months Post-operative	Ultrasound Biomicroscopy (UBM) performed, revealing normal ciliary processes and absence of anterior segment anomalies.
4 Months Post-operative	Best Corrected Visual Acuity (BCVA) improved to 20/50. Visual axis remained clear with good IOL stability.

DISCUSSION

We have presented the case of a male preschooler with a unilateral left pediatric cataract associated with esotropia. Notably, the patient lacked typical signs of persistent fetal vasculature (PFV) on clinical and sonographic examination, such as a fibrovascular stalk, microphthalmia, or ciliary body abnormalities. However, intraoperatively, a fibrovascular membrane tightly adherent to the posterior capsule was discovered.

Three strong differentials for this fibrovascular membrane with an associated cataract are cyclitic membranes, posterior capsular defects (PCDs), and anterior PFVs.

Cyclitic membranes are fibrovascular extensions from the ciliary body to the peripheral retina, often seen as complications of pars planitis.[12] However, the absence of uveitic history (floaters, pain) or signs (keratic precipitates, synechiae, vitritis), combined with the intralenticular location and normal postoperative UBM, makes this diagnosis unlikely.

Posterior capsular defects are rare forms of incomplete PFV regression characterized by thick defects with white dots.[6] While the intralenticular location and posterior capsule adherence are similar, the distinct fibrovascular structure observed in our case is a major point against this diagnosis.

Anterior PFV, defined by Anteby et al. as a cataract combined with a fibrovascular membrane, ciliary traction, or microphthalmia, remains the most likely diagnosis.[2,7] Pathologically, anterior PFV is usually characterized by retrolental fibrovascular membranes extending to the ciliary body, causing ciliary process elongation.[2] However, our case presented with atypical features: the fibrovascular membrane was located intralenticularly, and postoperative

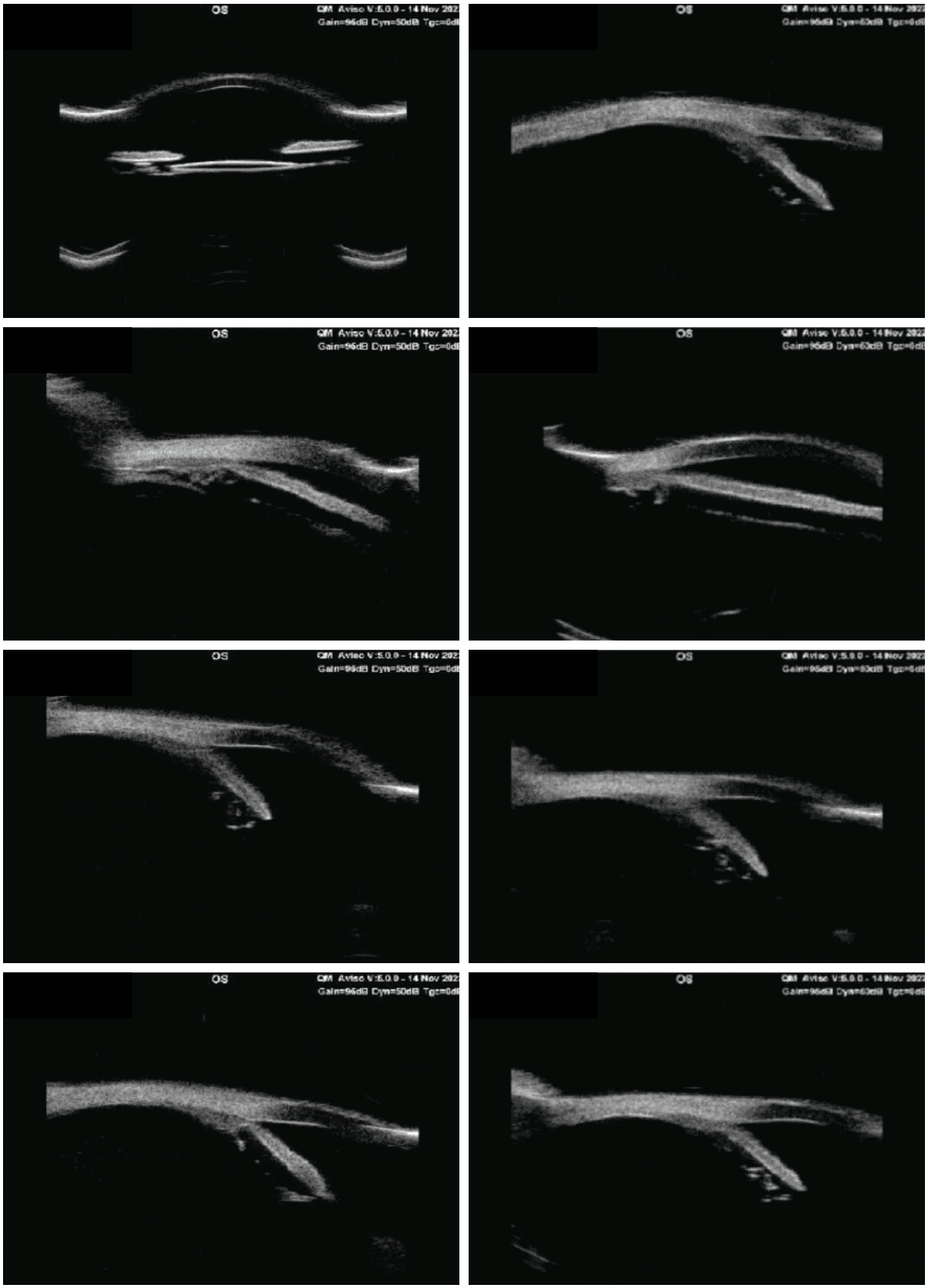

Figure 7. Ultrasound biomicroscopy of the left eye showed absence of elongation, incorrect position, and sources of neovascularization from the ciliary body and processes.

UBM confirmed the absence of ciliary process elongation or anterior segment abnormalities.

These atypical features can be explained by two mechanisms. First, the intralenticular location results from the extension of proliferation from the posterior capsule into the lens itself, contributing to cataract formation and potential intralenticular hemorrhage.[2] Second, the absence of ciliary body connections and elongation likely results from the continued regression of the fetal vasculature.[6] This regression is theorized to occur even after the pathology has developed, leaving no evidence of the initial ciliary involvement, as observed in other atypical PFV presentations.[6]

Since cataracts often obscure clinical examination, ancillary diagnostics are crucial.[8] In our case, the postoperative UBM confirmed normal ciliary processes, suggesting that the vasculature invading the ciliary processes may have regressed further following the tamponade effect of IOL implantation. Routine preoperative UBM is recommended for unilateral sporadic cataracts to detect such anomalies, as missed features can complicate surgery.[8]

Surgical management of PFV carries higher risks of hemorrhage and retinal detachment.[10,13] In this case, we employed partial excision of the membrane rather than complete peeling to minimize bleeding from the unknown vascular source. The absence of peripheral retinal attachments fortunately prevented retinal detachment. IOL implantation was successfully performed to prevent anisometropia, given the absence of microphthalmia.[10]

Postoperative vision improved in our patient, consistent with general outcomes for anterior PFV.[10] However, long-term monitoring for complications like visual axis obscuration and glaucoma, alongside aggressive amblyopia therapy, remains essential.

The primary strength of this case lies in the comprehensive documentation of the clinical course, from the initial consultation through intraoperative discovery to postoperative recovery, supported by complete multimodal imaging. However, a notable limitation is the timing of the ultrasound biomicroscopy (UBM). As this diagnostic was performed postoperatively, we cannot definitively rule out that the observed absence of ciliary process elongation was influenced by the anatomical changes following lens extraction and IOL implantation, rather than being a primary atypical feature.

With the wide spectrum of PFV as well as with the possible intraoperative and postoperative complications related to their cataract surgeries, the presence of these structures, including their atypical presentations, should be ruled out as part of pediatric cataract preoperative planning. This is important not only in surgical procedure planning but also in the prognostication in terms of visual outcomes, with the additional complications attributed to these cases.

Patient Perspective

The patient's mother reported significant relief regarding the resolution of the leukocoria, which had been a major source of concern for the family. However, she acknowledged that the current visual acuity does not represent the final outcome. Understanding the long-term nature of visual rehabilitation, she expressed a strong resolve to strictly adhere to the prescribed postoperative care and amblyopia therapy to maximize her child's visual potential.

CONCLUSION

This report presented a rare case of anterior PFV manifesting with an intralenticular fibrovascular membrane and a complete absence of ciliary process elongation. These atypical findings underscore the variable spectrum of the disease, supporting the theory of continued vascular regression even after pathological development. Consequently, we recommend that anterior PFV be routinely considered in the differential diagnosis of unilateral sporadic pediatric cataracts, even in the absence of typical clinical or sonographic signs. The utilization of high-resolution imaging, such as ultrasound biomicroscopy, is essential to detect these subtle anomalies, ensuring appropriate surgical planning and anticipation of potential intraoperative complications.

Informed Consent

The authors certify that the patient's mother has read, signed, and possesses a copy of the Informed Consent Form (ICF) regarding the publication of this case report and its accompanying images.

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Statement of Authorship

Both authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

Both authors declared no conflicts of interest.

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