Intriguing Interplay: A Rare Case of Persistent Pupillary Membrane Accompanying Funnel-Shaped Retinal Detachment

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ABSTRACT:
Persistent pupillary membrane (PPM) is a congenital ocular anomaly that typically manifests as fine strands of tissue across the pupil, remnants of a fetal vascular network. While most cases of PPM are benign and asymptomatic, rare instances can present with dense membranes that obstruct vision and lead to significant visual impairment. This report discusses the case of a 9-year-old male with unilateral dense PPM in the right eye, resulting in severe visual impairment. A thorough examination revealed normal corneal structure, but ultrasound B-scan showed a funnel-shaped retinal detachment. Given the retinal findings, surgical intervention for PPM was deemed inappropriate, and conservative management was chosen to minimize risks and preserve the patient's remaining vision. This case highlights the importance of individualized management strategies in complex presentations of PPM, especially when associated with additional ocular pathologies.

Introduction:

Persistent pupillary membrane (PPM) is a frequent congenital ocular anomaly, present in the majority of neonates (approximately 95%) and persisting in around 20% of adults [1]. It manifests as fine strands of tissue across the pupil, remnants of the anterior tunica vasculosa lentis, which plays a vital role in supplying nutrients to the developing lens during the first six months of fetal life [2,3]. Typically, these strands do not cause visual impairment and are considered a benign feature unless they persist in more significant forms or affect the visual axis.

In most cases, PPM consists of delicate fibers that do not require intervention. However, in rare instances, it may present as a total or dense membrane covering a significant portion of the pupil, potentially obstructing vision [4]. This can lead to visual disturbances and may necessitate clinical intervention. While the anomaly usually regresses or remains asymptomatic, some patients may experience complications like reduced visual acuity or amblyopia if the membrane obstructs light from entering the eye.

Case:

A 9-year-old male patient presented with a complaint of diminished vision in his right eye since birth. The family noted that the child had a history of visual impairment, but the severity of the condition had only recently become apparent to them. Upon evaluation, it was revealed that the patient's right eye exhibited a visual acuity of only perception of light (PL) positive, while the left eye demonstrated normal visual acuity of 6/6

A thorough examination of the extraocular muscle movements showed normal function, indicating no restrictions in eye movement. Anterior segment examination of the right eye was performed using a slit lamp, which revealed a clear cornea and a normally reacting pupillary margin. However, dense strands of tissue were observed attached to the iris collarette, extending across the pupil, consistent with a diagnosis of unilateral dense persistent pupillary membrane (PPM). The presence of these strands significantly obscured the pupil and was likely responsible for the patient's reduced visual acuity (Figure 1).

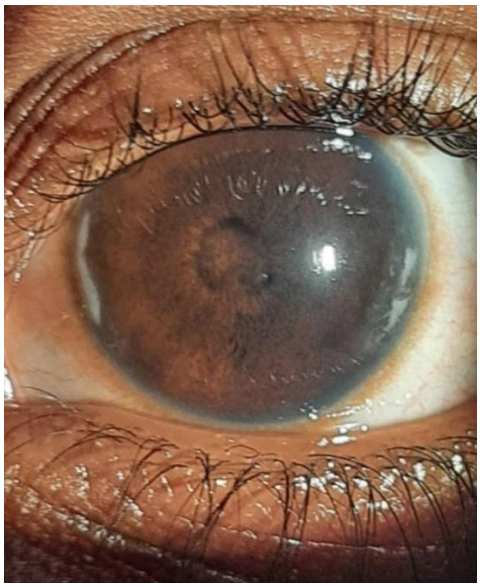

Figure 1: Clinical picture

To assess the integrity of the posterior segment, an ultrasound B-scan of the right eye was performed, revealing a funnel-shaped retinal detachment (Figure 2). Given this finding, surgical intervention such as membranectomy was deemed inappropriate, and conservative management was considered instead. A posterior segment examination could not be conducted, limiting the ability to assess potential complications in the retinal structures.

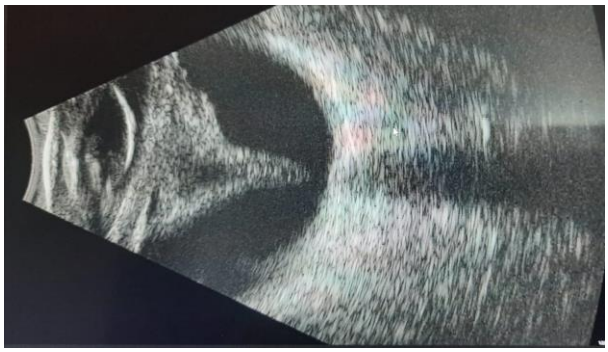

Figure 2: Ultrasound-B indicating funnel shaped retinal detachment

To further investigate the condition, specular microscopy was performed to rule out irido corneal pathology associated with anterior segment abnormality, providing insights into the corneal endothelium and anterior segment morphology. Specular microscopy of the right eye was conducted. The central corneal thickness (CCT) was 558 µm, which is within the normal range, indicating a structurally intact cornea. The endothelial cell density (CD) measured 3489 cells/mm², suggesting a healthy endothelium. Additional parameters, including the coefficient of variation (CV) of 22 and hexagonality (HEX) of 74%, further confirmed the corneal endothelium was normal. These findings suggested that corneal pathology was unlikely to be the primary cause of the patient's visual impairment. Given this, the differential diagnosis favored the dense persistent pupillary membrane (PPM) as the main contributor to the reduced vision, along with possible associated conditions such as sensory exotropia and chronic retinal detachment.

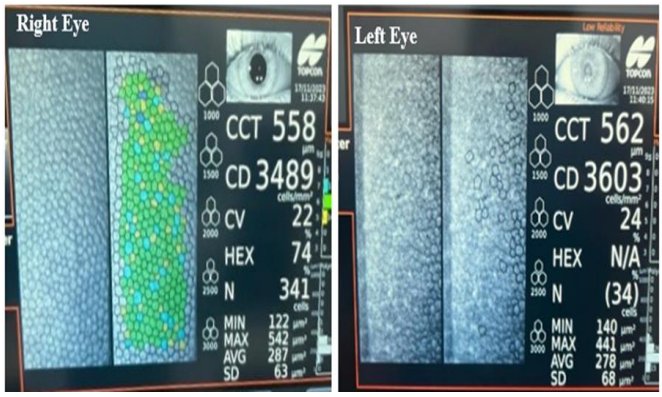

Figure 3: Specular microscopy

Given that only one eye was functional, a conservative management approach was chosen to preserve vision and

minimize the risks associated with surgery. This case illustrates the complexities of managing congenital ocular conditions like persistent pupillary membrane (PPM), highlighting the importance of a patient-specific treatment plan.

Discussion:

Persistent pupillary membrane (PPM) is a common congenital ocular anomaly, characterized by the presence of fine strands of tissue that extend across the pupil [1]. These strands are remnants of the anterior tunica vasculosa lentis, a vascular network that plays a critical role in supplying nutrients to the developing lens during the first six months of fetal life. Normally, the tunica vasculosa lentis regresses before birth, but in cases of PPM, parts of it remain attached to the iris collarette, forming these thin strands [2,3].

In most cases, PPM is benign and asymptomatic, consisting of delicate, non-obstructive fibers that do not interfere with vision. However, in rare instances, the membrane may be more extensive or dense, covering a large portion of the pupil and potentially obstructing light from entering the eye [5]. When this occurs, PPM can contribute to visual impairment, such as reduced visual acuity or sometimes complete loss.

As described in the by Banigallapati et al., 2018, when dense PPM obstructs the visual axis, early surgical intervention, such as membranectomy, may be required to restore vision and prevent long-term deficits [1]. However, in our case, the presence of a funnel-shaped retinal detachment discovered on ultrasound B-scan contraindicated surgical intervention due to the heightened risk of complications. Studies have also shown that surgical interventions in eyes with compromised posterior segment structures may exacerbate the risk of vision loss and other complications [6,7].

Given that the patient's left eye had normal visual acuity, conservative management was preferred to avoid the risks associated with surgery. This decision aligns with the literature, which emphasizes individualized treatment based on the extent of the obstruction and the overall visual prognosis [8–10]. Conservative approaches may be more appropriate in cases where there is limited visual potential in the affected eye, and the unaffected eye is fully functional [10]. This case demonstrates the need for a careful, patient-specific approach in managing complex presentations of PPM, especially when associated with other ocular pathologies, such as retinal detachment.

Conclusion:

his approach highlights the importance of individualized treatment plans, taking into account both the severity of the membrane and the overall visual prognosis. When managing cases of PPM, particularly in the presence of other ocular pathologies, careful consideration of the risks and benefits of surgical versus conservative treatment is crucial to preserving vision and minimizing complications.

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