Oral NSAIDs are the first line of treatment for PS; however, immunosuppressive drugs are often needed to fully control the inflammation.[2,6] One study reported on 20 eyes of 13 pediatric patients with PS and reported flare-ups when tapering the oral corticosteroid to <0.5 mg/kg/day.[2] Owing to exhibiting side effects or relapse on low-dose corticosteroid, at least one immunosuppressive drug was added to control the inflammation. In the current case report, the patient experienced 3 separate episodes of flare-up whilst on full dose of methotrexate and mycophenolate when oral prednisolone was reduced to 10 mg/day (0.2 mg/kg/day). Therefore, infliximab 300 mg every 4 weeks was successfully started to control his inflammation. Only one previous case report on paediatric PS used infliximab for disease resistant to methotrexate, methylprednisolone, and cyclosporine treatment.[10] Weiss et al. presented the case of a 13-year-old girl diagnosed with PS who maintained remission with 10 infusions of infliximab (5 mg/kg) with no side effects over a 16-month follow-up.[10] Our case likely represents a more severe case requiring much longer immunomodulation with infliximab (5 years total) to achieve long-term remission. Although there are published data on infliximab therapy for childhood refractory uveitis,[11–13] it is encouraging to report safe and effective disease control in PS as well.

In conclusion, the majority of pediatric PS cases in literature report poor disease control with only NSAIDs or corticosteroids, with many requiring at least one immunosuppressive agent to avoid steroid side effects. We propose that infliximab can be considered an alternative treatment for severe paediatric PS resistant to treatment.

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Iris stromal defect in an infant masquerading as a tumour: the sailing iris?

Congenital iris lesions are rare and generally identified early in life.[1,2] A pigmentary lesion on the iris ranges from a simple nevus or cyst to tumors including melanoma. These lesions can arise from both iris stroma and iris epithelium.[2] Thorough examination as well as anterior segment imaging studies may be required to establish a diagnosis.[3] Here, we report a rare case of congenital iris stromal defect masquerading as an iris tumour.

Methods

The history, systemic and ocular findings, and investigations of a patient with a congenital iris lesion was reviewed. Written consent was obtained from the patient's guardian.

Case report

A 3-month-old boy with no significant birth or family history, presented with an abnormality of the left iris. On slit lamp examination, a pigmented iris lesion in the

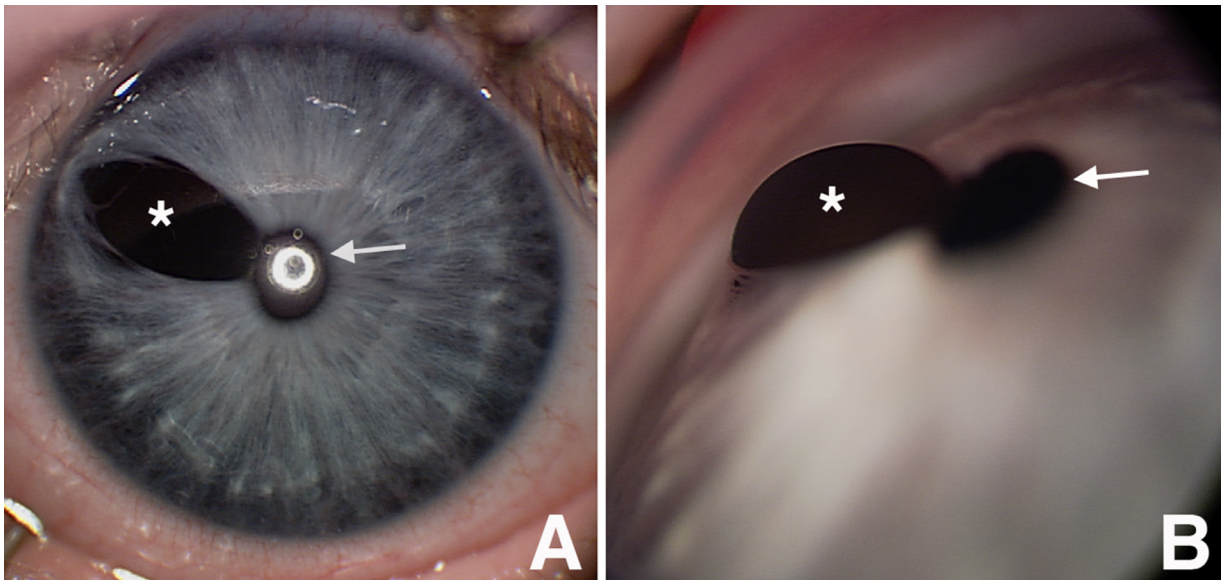

Fig. 1—Magnified photo of iris. (A) The iris with the pigmented lesion (*) at the supranasal quadrant identified as elevated from the surrounding normal iris. The arrow shows the nondilated pupil and ring flash reflex. (B) A gonioscopic view of how the lesion (*) is elevated above the iris plane. The pupil is marked with an arrow.

supranasal quadrant of the iris was noted. The lesion had a smooth surface, appeared solid, measured $3.5 \times 2 \times 1$ mm in size, and did not distort the pupil (Fig. 1). The child had fix and follow vision, normal ocular motility and intraocular pressure. Ultrasound biomicroscopy was performed before pupillary dilation, which showed absence of the anterior iris stroma with bowing forward of the posterior pigmented epithelium of the iris without a cyst wall or solid component (Fig. 2A). Upon pupillary dilation, the pigmented epithelium no longer bulged forward and collapsed to a wrinkled sheet (Fig. 2B). The rest of the eye exam, including gonioscopy, lens, and fundus, was normal in both eyes. All other investigations including optical coherence tomography of the fovea and genetic testing for PAX6 mutations were normal. A diagnosis of isolated congenital absence of iris stroma was made. He was followed for 7 years during which the defect remained stable with no changes or evidence of ocular pathology. He was diagnosed with autism during childhood. At final follow-up,

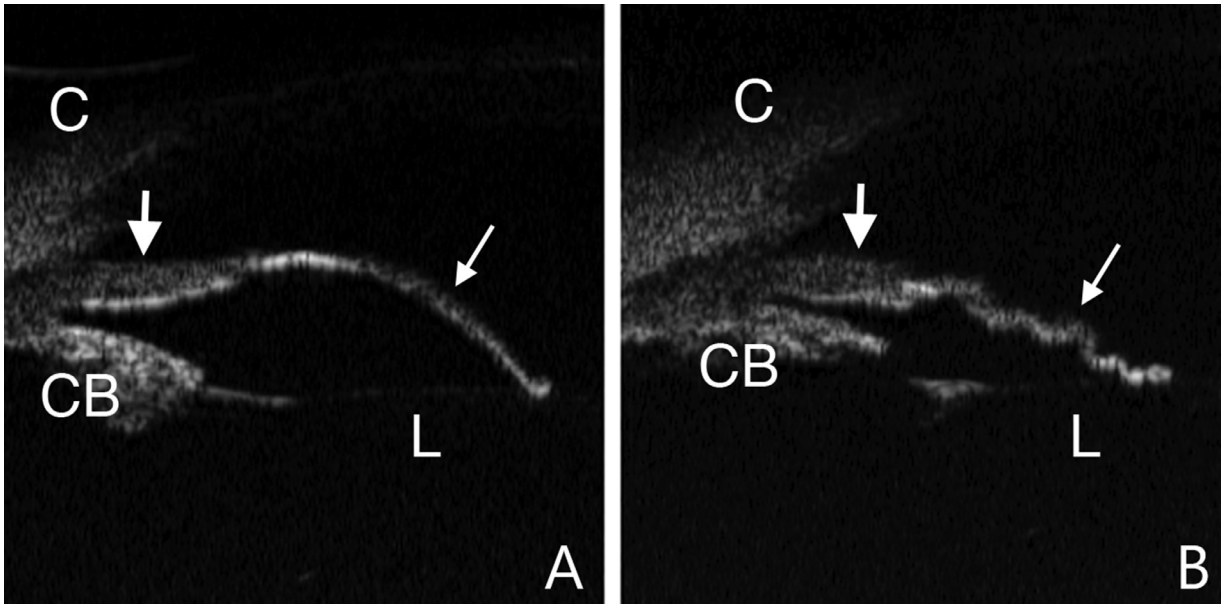

Fig. 2—Ultrasound biomicroscopy showing the cornea (C), the ciliary body (CB), and the lens (L). The thick arrows demonstrate the normal iris stromal thickness in the periphery. The thin arrows show the thin posterior pigmented epithelium without overlying stroma. (A) The thin iris bowing forward when the pupil is not dilated. (B) The flaccid iris after pupillary dilation. Note the absence of a cyst wall.

both eyes had visual acuity of 0.0 logMAR with normal intraocular pressure and healthy fundus.

Discussion

This case of isolated congenital iris stromal defect can be explained by understanding the relationship between intraocular fluid dynamics and the structure of the iris. Forward flow of aqueous from the posterior chamber to the anterior chamber is driven by a pressure gradient across the pupil, with pupillary diameter being a main factor affecting the magnitude of this gradient.[4] As the pupil constricts, the iris is in greater apposition to the lens, increasing this pressure gradient. Conversely, a dilated pupil results in a decreased pressure gradient across the chambers.[4]

In the present case, the absence of stroma in the abnormal portion of the iris resulted in a thin and floppy structure that was easily distended by physiologic pressure gradients. When the pupil was small, the relatively high physiologic pressure gradient behind the iris led to a forward bowing of the posterior pigmented epithelium. The physics of this is analogous to that of a sail in the wind. When the pressure generated on the posterior surface of the sail is greater than that of the anterior surface, the sail bows forward. When the pupil dilated and the pressure gradient decreased, the posterior pigmented epithelium resumed a flaccid position and the apparent "mass lesion" disappeared. This is analogous to when there is no wind in a sail, hence, the sailing iris!

Other congenital iris defects are described in the literature including colobomas, aniridia, Axenfeld-Rieger, and Irido-corneo-endothelial syndromes.[5] These defects do not result in the same "sailing iris" effect, as no pressure gradient is created due to either full thickness iris defects or a lack of pupillary contact with the lens. In contrast, the consequences of pressure gradients between anterior and posterior of the iris are observed in the elderly. With the development of cataract, the lens thickens and pushes forward against the pupil. This creates a pressure gradient that bows the entire peripheral iris forward, even in the presence of normal iris stroma, leading to acute angle closure glaucoma in predisposed eyes with shallow iridocorneal angles.[6] In the present case, the local bowing of the iris has not led to any angle closure or pressure rises over a 7-year follow-up. This case demonstrates that detailed examination, under anesthesia as required, including imaging modalities are essential to assessment of pigmentary iris lesions.

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Conjunctival and periorbital petechiae presumed secondary to self-inflicted asphyxiation in a pediatric patient

The presence of petechiae of the face and conjunctiva is a well-described finding seen in strangulation injuries. These petechiae are commonly seen post-mortem in deaths by asphyxia and are consequently used in forensic analysis.[1] Conjunctival and facial petechiae have also been noted to occur in nonfatal asphyxia, and their presence may be a sign that a severe strangulation injury has occurred.[2] The mechanism for facial and conjunctival petechial hemorrhages in asphyxia is thought to be rupture of capillaries with little connective tissue support secondary to local pressure elevation from continued arterial flow while venous output is obstructed.[1] We herein report an unusual case of a pediatric patient in which this clinical presentation signaled underlying recreational self-inflicted strangulation. This case report is adherent to the principles of the Declaration of Helsinki and is compliant with Health Insurance Portability and Accountability Act guidelines.

A 13-year-old girl presented with conjunctival and periorbital petechiae, noticed several hours earlier. She denied change in vision, eye pain, nosebleeds, or easy bruising. She had a history of chronic constipation but no recent infections, weight change, or fever and no family history of bleeding disorders. When interviewed in private, the