Sir,

Plateau iris syndrome in a child

Plateau iris is an angle appearance in which a short iris root angulates forward and centrally, and the iris surface is flat.¹ It occurs most commonly in younger women and tends to be hereditary.² The youngest previously reported patient was 22 years old.³ Rare cases of primary angle-closure in children have been reported, but none had plateau iris,^{4–7} making this, to our knowledge, the youngest patient reported.

Case report

A 12-year-old Hispanic boy presented with transient eye discomfort both eyes (OU), while watching television in the dark. Uncorrected visual acuity was 20/20 right eye (OD) (+0.50–0.25 × 95), 20/25 OS (+1.50–0.50 × 53). Intraocular pressures were 17 mmHg OD and 15 mmHg left eye (OS). Dark room gonioscopy revealed a convex iris approach with appositional closure OU and a double hump sign. Indentation gonioscopy opened the angle 360 degrees without peripheral anterior synechiae. Optic discs and visual fields were normal. Ultrasound

biomicroscopy showed an enlarged ciliary body displacing the peripheral iris forward, with loss of the ciliary sulcus (Figure 1). After laser iridotomies, the angles remained appositionally closed. A second ultrasound biomicroscopy (UBM) confirmed the gonioscopic findings (Figure 2). Argon laser peripheral iridoplasty was performed successfully to eliminate the plateau iris component of the angle closure.

Comment

Anatomically, plateau iris is characterized by a relatively flat iris plane, large, anteriorly positioned ciliary processes, absence of the ciliary sulcus, a sharp and angulated drop-off of the peripheral iris, and narrowing or closure of the iridocorneal angle.^{8,9} Plateau iris syndrome is defined by continued or inducible angle-closure in an eye with plateau iris configuration, following the elimination of pupillary block by iridotomy.⁵ The ciliary processes prevent the iris root from falling posteriorly. The age of presentation is determined by several factors including the extent of anterior displacement and the continued growth

Figure 1 UBM depicting appositional angle closure before laser peripheral iridotomy with angulations of peripheral iris (white arrows) and deep appearing anterior chamber. Enlarged ciliary body observed displacing peripheral iris forward (star) with loss of ciliary sulcus. Black arrow indicates scleral spur.

Figure 2 UBM of the angle after peripheral iridotomy of both eyes show continued appositional angle closure (arrows). Black arrow indicates scleral spur.

of the lens contributing to progressive angle crowding.¹⁰

Peripheral iridotomy eliminates pupillary block and argon laser peripheral iridoplasty addresses the residual angle closure.¹¹ Patients with appositional angle closure may go on to develop peripheral anterior synechiae (PAS) and synechial angle closure, even years after a successful iridotomy. Patients who underwent iridotomy should not be considered cured but should undergo periodic gonioscopic examination.

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Sir,

Comment on 'A PAX6 gene polymorphism is associated with genetic predisposition to extreme myopia'

Tsai *et al*¹ carried out a case–control association study in 340 Taiwanese medical students, and reported an association between presence of 'extreme myopia' (< −10.00 D) and genotype at SNP rs667773 (also known as IVS9-12C to T) in the PAX6 gene. No association was found between 'high myopia' (< −6.00 D) and rs667773 genotype.

rs667773 has a minor allele frequency (MAF) 4% in the United Kingdom; 7% in CEPH Europeans; 11% in Japan, and has been suggested to be a neutral polymorphism unlikely to cause an overt phenotype such as aniridia.^{2,3} No common (neutral) PAX6 variants have been associated with myopia susceptibility.^{4,5} We suggest that until replicated, the findings of Tsai *et al* should be interpreted with caution for the following reasons.

First, the sample size for the subgroup analysis was limited ($n = 67$ extreme myopes; 85 controls). Thus, the risk of a type I error was high. The moderate heterozygosity of rs667773 exacerbates this difficulty (hence the very low counts of TT genotypes).

Second, the issue of population stratification was not considered. The MAF of rs667773 in the study of Tsai *et al* was 17%, suggesting that T allele is more common in Taiwanese than in hitherto studied populations. This wide variability in allele frequency could easily lead to a false–positive association if refractive error also varied across the region from which the subjects were sampled.

Third, we note that the higher-risk C allele is more common in Europeans than in East Asians, whereas extreme myopia is more common in East Asia than in Europe. If rs667773 is an important myopia susceptibility variant, one would expect the opposite relationship.

Fourth, according to the common disease, common variant hypothesis, for a high-risk variant to rise to a frequency >80%, it would have to exert a strong selective advantage. No such advantage has been attributed to the C allele of rs667773.

Finally, unless the genetic risk factors for high and extreme myopia are distinct, the lack of association with high myopia is unexpected, especially since power was greater to detect an association with high myopia than extreme myopia.

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