Ruptured unilateral anterior lenticonus with congenital hypertrophy of retinal pigment epithelium: A rare association

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Key words: Alport, congenital hypertrophy of retinal pigment epithelial lesion, lenticonus

A 17-year-old boy presented to our hospital with complaints of defective vision of the left eye (OS) since 3 weeks. The patient gave a history of fall of chalk powder in OS 1 month back.

Best-corrected visual acuity was 20/40 in the right eye (OD) and 20/1200 in OS. Slit lamp biomicroscopy revealed OD anterior lenticonus [Fig. 1] and OS ruptured anterior lens capsule [Fig. 2] with dense heterogeneous lenticular opacity in a circular pattern [Fig. 3]. Fundoscopy OD showed Congenital Hypertrophy of Retinal Pigment Epithelium (CHRPE) [Fig. 4] and OS was hazy, with normal B scan. Systemic evaluation revealed microcystic hematuria and bilateral moderate sensorineural hearing loss. He underwent OS cataract extraction with IOL implantation. On follow-up, till 4 months the patient is maintaining the best-corrected visual acuity of 20/30. He has been started on ACE inhibitors by the nephrologist.

Figure 1: Slit lamp biomicroscopy image revealing the right eye anterior lenticonus

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Discussion

Anterior lenticonus is predominantly X-linked inherited (85%).[1] It is more common in males.[2] The pathogenesis of Alport's syndrome is explained by loss of collagen IV α3α4α5 network in basement membranes of the eye.[3] The absence of α3α4α5 network from the capsule leads to partial splits that may rupture with minor trauma or even spontaneously.

The most frequent ocular findings in Alport's syndrome are lenticular and retinal abnormalities, which include anterior lenticonus and fleck retinopathy. Other rare findings include posterior polymorphous corneal dystrophy, posterior lenticonus, cataract, and temporal macular thinning.[1,4] From the literature review, we found that boys in adolescent age are at particular risk for spontaneous lenticonus rupture and a protective eyewear is recommended to avoid any trivial trauma. We also found a rare association of CHRPE lesion, which has not been reported earlier. This finding prompted a need for endoscopic evaluation to rule out Familial Adenomatous Polyposis (FAP).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have

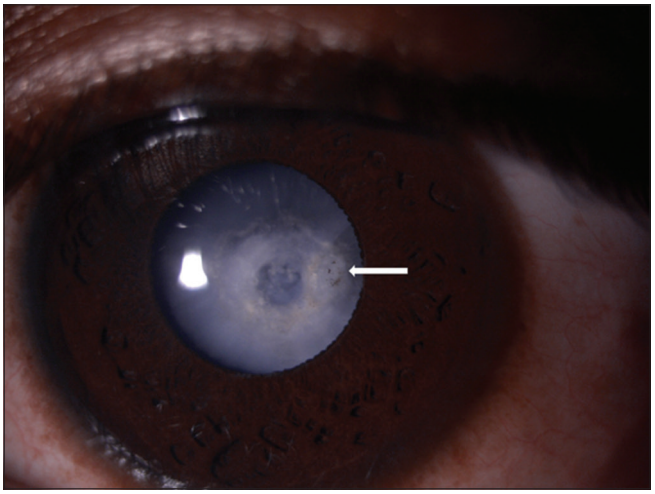

Figure 2: Slit lamp image of the left eye showing ruptured anterior lens capsule and localised underlying cataract

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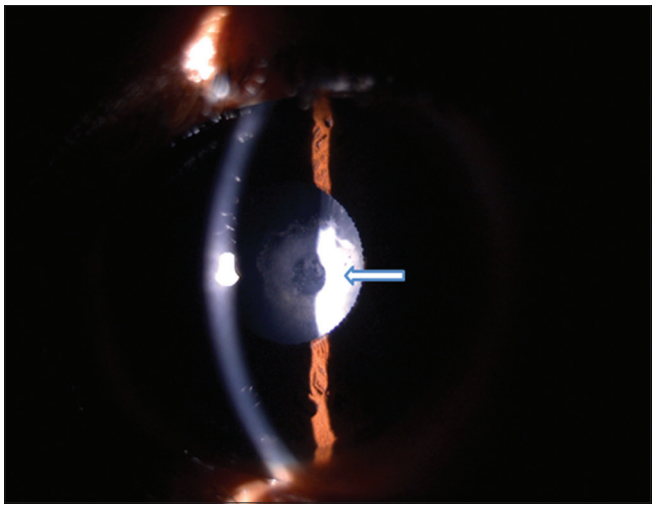

Figure 3: Slit lamp photo of the left eye showing dense heterogeneous lenticular opacity in a circular pattern

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

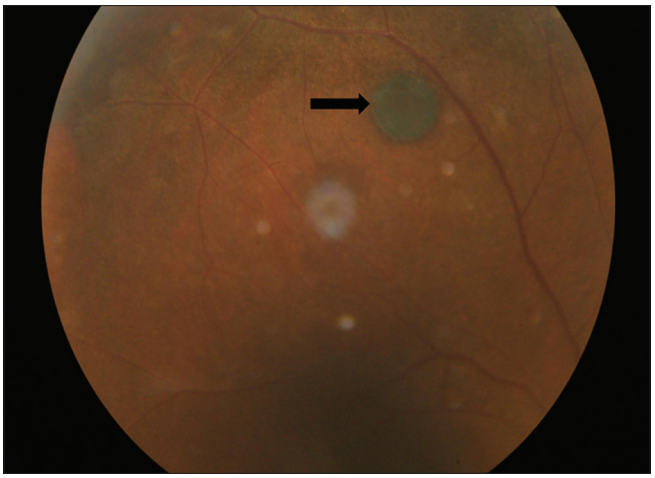

Figure 4: Fundoscopy of the right eye revealing Congenital Hypertrophy of Retinal Pigment Epithelial lesion (CHRPE) lesion

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