

Bilateral lens subluxation in a case of Behcet's disease related uveitis

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Ocular inflammation accompanying the pediatric Behcet's disease (BD) is characterized by a relapsing and complicated course, often resulting in blindness due to severe bilateral affection (1). Ectopia lentis (EL) is an important association of multiple ocular and systemic syndromes, with favorable visual outcomes if managed ideally (2). We discuss a unique case presenting with a combination of these entities, and explore the possible causes.

A 20-year-old male presented to our clinic with severe vision loss since 10–12 years of age, along with history of recurrent redness and pain in early childhood that was managed with topical and oral steroids as per old documents. There were definite history and signs of multiple aphthosis; oral as well as genital; along with positivity for HLA B5, but no significant family history or mental disability. The vision loss had become more severe in the past few years with present corrected visual acuity being hand motions close to face OU. OU subluxation of the lens was noted, OS having dense absorbed cataract with capsular fibrosis while OD had complicated posterior subcapsular cataract (*Figure 1*). There were also signs of previous panuveitis OU, with few old keratic precipitates in both the eyes accompanied by severe sclerosing mixed retinal vasculitis, maculopathy and optic atrophy (*Figure 2*). Intra ocular pressure (IOP) was normal OU. Serum Treponemal antibody tests were negative and urine examination was negative for amino aciduria. Marfan's syndrome was ruled out with Echocardiography. The patient was diagnosed BD after an immunology consultation. OS lens extraction was performed under explained prognosis.

The vision improved minimally after the surgery to counting fingers close to face, and remained stable for

2 years when he again presented in the emergency with sudden onset painful vision loss OD. He had developed phacotopic glaucoma, with complete anterior dislocation of lens leading to IOP of 60 mmHg (*Figure 1*). He underwent an urgent lens extraction OD, and corrected visual acuity improved to counting fingers close to face. Three months hence, condition is stable OU with stable vision, normal IOP and quiescent uveitis.

Pediatric BD are known to commonly present as panuveitis with retinal vasculitis, having poor visual prognoses and high odds of ocular complications like optic atrophy, especially in presence of HLA B5 positivity (1). While the diagnosis of BD was straightforward as per the International criteria (3), we could not identify any known cause of EL. EL in our case appears to be a complication of the ocular inflammation. Spontaneous lens subluxation is a known end result of long standing uveitis and has been described with Vogt Koyanagi Harada syndrome, sarcoidosis and pars planitis (4). Hence accumulation of end products of inflammation in a closed compartment like the eye ball may result in zonular weakness, especially due to the proximity of the zonules and the zonular girdle to the uvea. In fact, late post-operative bag—IOL dislocations have also been seen in eyes having uveitis (5). Accumulation of inflammatory end products over the zonules and the lens capsule has been seen in cases of BD histo-pathologically too (6). EL, therefore, could have occurred due a combination of capsular shrinkage as well zonular dehiscence. Because of different genetics (3) and uncommon targets of inflammation (3,7), direct association between BD and EL is unlikely. Also the unsynchronized occurrence of ocular BD and EL in our case renders this association unlikely.

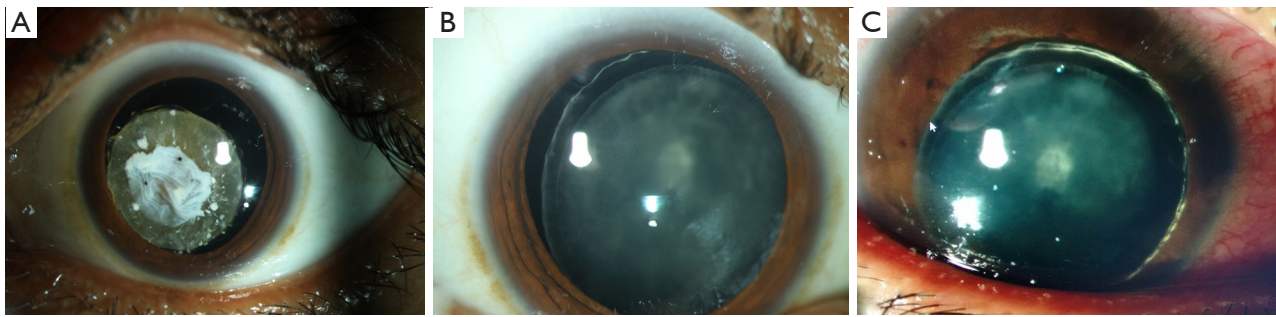


Figure 1 Clinical photographs of the anterior segment. (A) Subluxated cataractous lens OS. Note the anterior capsular shrinkage/dense plaque with remnants of posterior synechiae; (B) subluxated lens OD with complicated cataract; (C) anteriorly dislocated cataractous lens OD noted after follow-up of 2 years. Note the conjunctival congestion due to acute glaucoma.

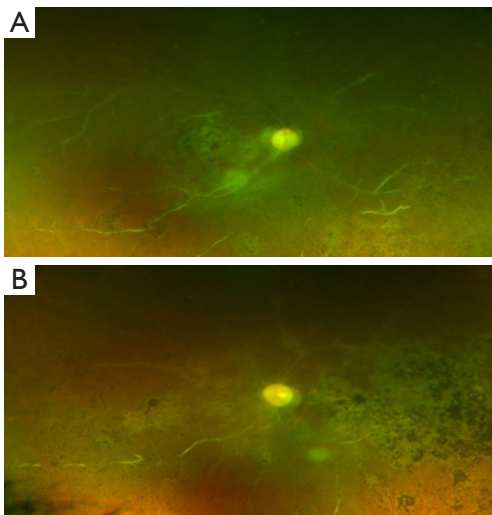


Figure 2 Ultra-wide fundus photograph OU depicting mixed vascular sclerosis, macular atrophy and optic atrophy. Pigmentary changes can also be seen throughout the fundus.

Hence, EL may occur as a complication of long standing panuveitis in cases of BD. The visual prognoses largely depend on functional status of the posterior segment.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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