Intercalary staphyloma in Marfan syndrome: A dreaded complication of scleral incision

Pranita Sahay, Suman Dhanda, Prafulla K Maharana, Jeewan S Titiyal

Key words: Ectopia lentis, intercalary staphyloma, intracapsular cataract extraction, Marfan syndrome, scleral incision

A 28-year-old male patient, known case of Marfan syndrome, presented with complaints of low vision in both eyes since childhood. He gave a clinical history of having undergone intracapsular cataract extraction (ICCE) in the right eye 3 years previously. At presentation, the best spectacle corrected visual acuity (BCVA) was 6/12 and 1/60 in the right eye and left eye, respectively. On slit lamp examination, the right eye had an intercalary staphyloma extending from 11 to 2 o' clock with Descemet scar in the superior cornea [Fig. 1]. The iris showed atrophic patches, a fixed and dilated pupil and aphakia with no capsular support [Fig. 2]. The left eye had iridodonesis with 8 clock hours of nasal subluxation of a clear crystalline lens [Fig. 3]. Fundus was noted to be normal on indirect ophthalmoscopy. Intralenticular lens aspiration was planned for the patient's left eye. However, the patient refused surgery.

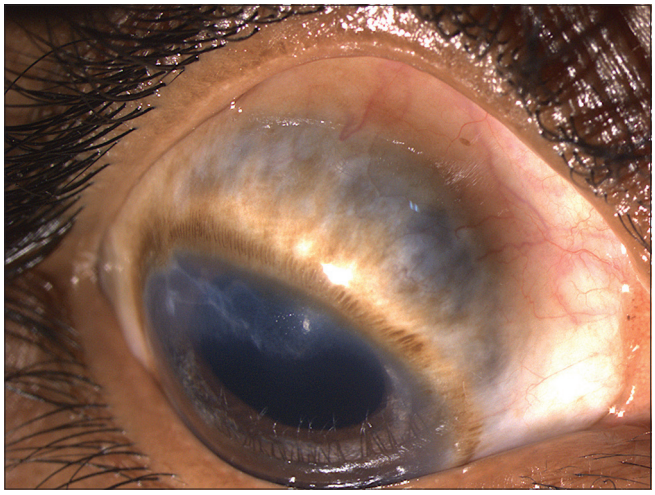

Figure 1: Slit lamp photograph of right eye showing an intercalary staphyloma, superior descemet scar, and aphakia

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Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence to: Dr. Prafulla K Maharana, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India. E-mail: drpraful13@gmail.com

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Discussion

Marfan syndrome is characterized by abnormalities of the fibrillin protein that is responsible for providing strength

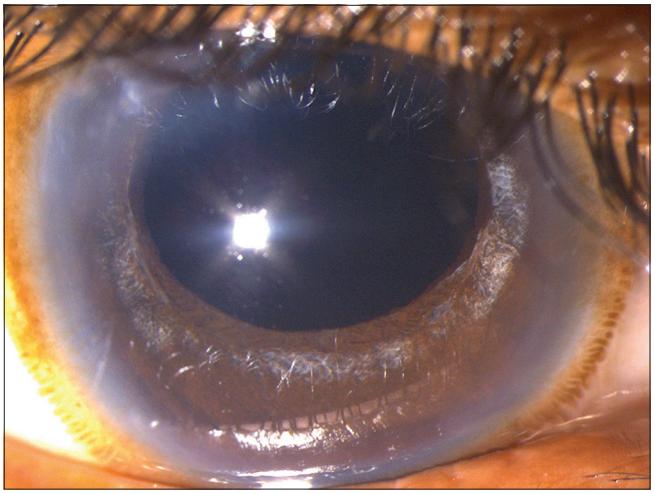

Figure 2: Slit lamp photograph of right eye showing 360° iris atrophy with aphakia

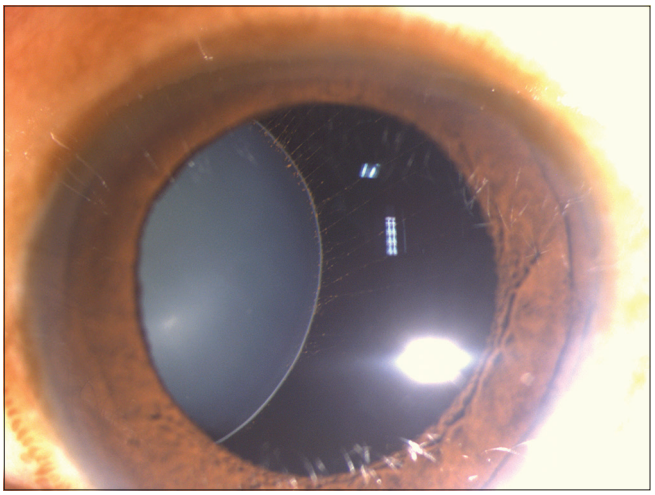

Figure 3: Slit lamp photograph of left eye showing nasally subluxated lens with superotemporal stretched zonules and inferotemporal deficient zonules

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and elasticity to the connective tissue.[1,2] "Ectopia lentis" is a common finding in Marfan syndrome that often requires surgery for visual rehabilitation.[3] The ectopic lens can be removed through a corneal, limbal, or scleral incision. However, there are reports in literature that suggest the risk of staphyloma formation following ocular trauma and scleral incisions in these cases.[4,5] Goldberg and Ryan[4] reported an intercalary staphyloma in a 5-year-old female child of Marfan syndrome following blunt ocular trauma with a stone. Seelenfreund *et al*.[5] reported occurrence of an incisional staphyloma following retinal detachment surgery in a case of Marfan syndrome. Therefore, this case highlights an important lesson that any form of trauma to the sclera (surgical or mechanical) should be avoided in cases of Marfan syndrome considering the pre-existing scleral thinning and risk of staphyloma formation.

Declaration of patient consent

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

There are no conflicts of interest.

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