A rare association of optic disc pit maculopathy and ectopia lentis

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Key words: Ectopia lentis, optical coherence tomography, optic disc pit maculopathy, optic disc pit

A 14-year-old boy presented with a sudden onset painless diminution of vision in the left eye for the past 2 months. There was no history of trauma or any systemic illness. Best-corrected visual acuity was 20/20 and 20/200 in his right and left eye, respectively. Slit-lamp examination of left eye revealed superonasal ectopia lentis with stretched zonules visible from 2 to 7 clock hours [Fig. 1a and b]. Complete physical examination and cardiac evaluation including echocardiography, tests of joint hyperflexibility, serum homocysteine levels were done to rule out any associated systemic disorders. Dilated fundus examination and fundus imaging (Optos T × 200, Optos PLC, Dunfermline, Scotland, UK)) of the left eye revealed an oval, greyish-yellow crater-like depression of size 1/10th the disc diameter at the temporal aspect of optic disc suggestive of an optic disc pit (ODP) associated with serous macular detachment [Fig. 2]. Swept-source optical coherence tomography (SSOCT-Triton-Topcon, Tokyo, Japan) of the macula revealed retinoschisis involving multiple layers, i.e., outer segments of photoreceptors, outer nuclear layer, and outer plexiform layer [Fig. 3a]; and neurosensory detachment in the left eye extending up to the temporal margin of the disc. [Fig. 3b] Patient underwent Pars plana vitrectomy with posterior vitreous detachment (PVD) induction with instillation of platelet-rich plasma at the site of disc pit.

Discussion

Optic disc pit maculopathy is a rare congenital anomaly. The pathogenesis of optic disc pit is still controversial, with one of the hypothesis describing it as an incomplete closure of the superior edge of the embryonic fissure.[1] Unilateral Ectopia lentis is another uncommon congenital disorder, due to defective

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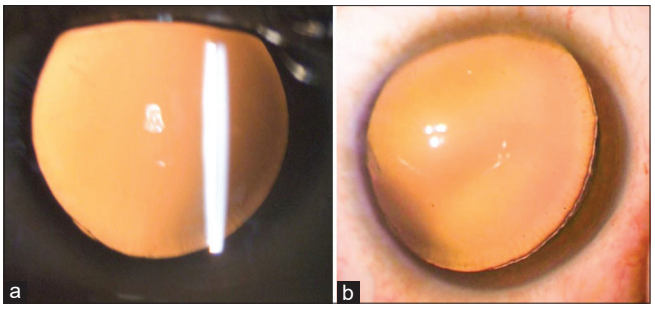

Figure 1: (a) Slit lamp photograph of normal right eye. (b) Slit-lamp photograph of left eye showing superonasalectopialentis with stretched zonules visible from 2 to 7 clock hours

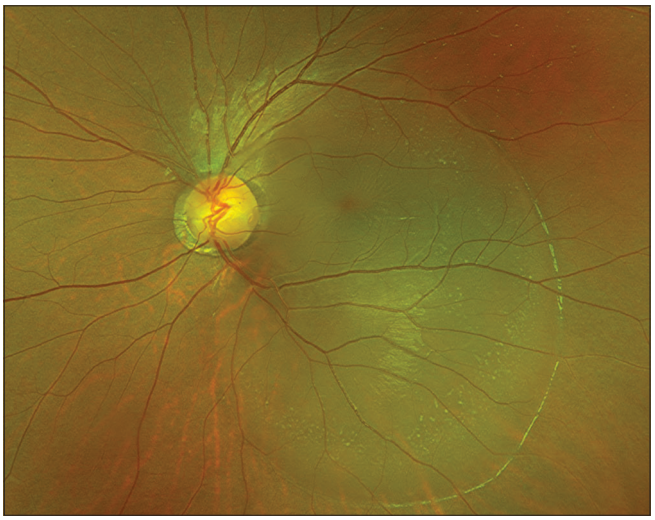

Figure 2: Color fundus photograph of left eye depicting optic disc pit and associated serous retinal detachment

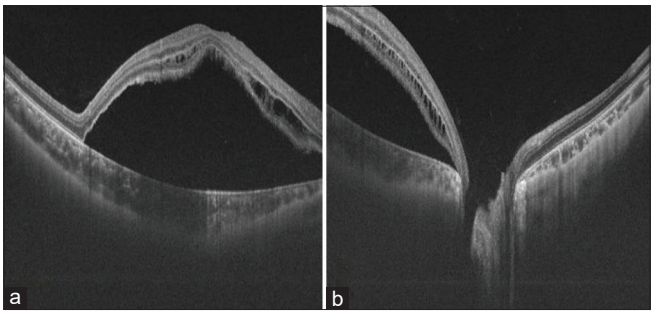

Figure 3: (a) Swept source OCT image of left eye showing neurosensory detachment at macula with retinoschisis involving multiple layers. (b) Swept source OCT image of left eye showing neurosensory detachment extending upto the margin of the disc

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zonular development.[2] The association of optic disc pit and ectopia lentis is peculiar. However, It has been found that lamina cibrosa is absent at the site of the optic pit.[3] It is noteworthy that both lamina cibrosa and zonules are made up of fibrillin-rich microfibrils.[4] However, no such association has been previously described. Comprehensive retinal examination with multimodal imaging techniques can help to arrive at the diagnosis.[5]

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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