Grand Rounds

A 37-year-old woman with spontaneous lens dislocation after vitrectomy for retinal reattachment

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History

A 37-year-old Afro-Caribbean woman with Marfan syndrome presented with a history of blurred vision in the right eye for 7 days. Her medical history included cardiomyopathy with congestive heart failure.

Examination

Visual acuity was 6/12 on the right and 6/6 on the left eye. She was phakic and there was no evidence of subluxation of the lens. She had a macula-on retinal detachment affecting the nasal and infero-temporal part of the retina and two superior retinal tears were found at 12 o'clock.

Ancillary Testing

No additional testing was performed.

Treatment

The patient underwent uncomplicated vitrectomy, cryotherapy and 16% C3F8 gas injection under local anaesthetic. The operation was uneventful and no lens instability was noted peri-operatively. Her visual acuity on the first postoperative day was hand movements, there was mild anterior chamber reaction and the intraocular pressure was 26 mmHg. The lens was stable but we noted some mild posterior subcapsular opacities due to the presence of gas. The retina was attached and there was 80% fill of the vitreous cavity with gas.

Two weeks later, the visual acuity was hand movements in the right eye. The lens had dislocated to the inferior vitreous cavity, with recurrence of the retinal detachment (Figure 1).

Figure 1. Lensectomy of the crystalline lens dislocated into the vitreous cavity.

The patient underwent repeat vitrectomy, lensectomy to remove the dislocated lens, cryotherapy to a dialysis at 6 o'clock and 16% C3F8 gas tamponade. On the first post-operative day visual acuity was hand movements, there was moderate corneal edema and 80% fill of the vitreous cavity with gas.

Two months post-operatively the best corrected visual acuity is 6/12 in the right eye with aphakic correction. The plan for the patient is to perform a secondary intraocular lens implantation. There was no lens instability of the fellow eye during the whole period that the patient was under our care.

Differential Diagnosis

The differential diagnosis includes zonular weakness due to Marfan syndrome and intraoperative zonular injury.

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Diagnosis

Our patient had a known diagnosis of Marfan syndrome in which lens dislocation due to zonular weakness is a common feature. The main constituent of the ciliary zonules that supports the lens is the glycoprotein fibrillin. It has been suggested that mutations in this glycoprotein are responsible for increased susceptibility to the proteolytic activity of serine proteases and matrix metalloproteinases, compromising the structural integrity of the zonules.¹

Well-known conditions that are associated with fibrillin disruption are Marfan syndrome and ectopia lentis and both may present with lens dislocation.¹ Previous reports have highlighted on surgery for subluxed/dislocated lens in Marfan patients.^{2,3} There is also reference in literature about retinal re-attachment surgery in such patients.⁴ Spontaneous lens dislocation has been reported in pseudoexfoliation,⁵ uveitis⁶ and following scleral buckling procedure.^{7,8}

In our patient the causes for the spontaneous dislocation of the lens could be attributed to either a subclinical zonular weakness related to Marfan or due to peri-operative imperceptible zonular damage during vitrectomy and subsequent gas pressure on the lens. Surgery for retinal

re-attachment has been associated with many postoperative complications. To the best of our knowledge spontaneous dislocation of lens following vitrectomy is a very rare complication.

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