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Sir,

Late spontaneous anterior dislocation of an intraocular lens (IOL) with the capsular bag

Spontaneous luxation of an intraocular lens (IOL) with the capsular bag after uneventful cataract surgery is rare. Predisposing factors have been described as: high myopia, psedoexfoliation,[1,2] uveitis, pars planitis, myotonic dystrophy, diabetes mellitus, increased age and retinitis pigmentosa.[1,3] To our knowledge, there have been only five such reports[4–8] wherein spontaneous luxation was attributed to trauma,[4] atopic dermatitis,[7] and capsular contraction.[5,6,8] The maximum time span was 9 years[5] after cataract surgery, and all were posterior dislocations.

We report a case of bilateral spontaneous dislocation of an IOL 14.5 and 9 years after cataract surgery as a presenting feature of possible Marfan's syndrome.

Case report

A 41-year old man presented with a day's history of sudden, painless blurring of vision in his left eye. Previous ophthalmic history included an uneventful phacoemulsification with IOL in the left eye (14.5 years before presentation), and an extracapsular cataract extraction with secondary sulcus fixated IOL followed by a retinal detachment 4 years later (treated with pars plana vitrectomy and silicon oil) in the right eye. Subsequently, the right eye developed a silicon keratopathy which was treated with a penetrating keratoplasty. He presented in September 2000 (9 years after cataract surgery) with a spontaneous posterior luxation of the PCIOL in the right eye, which was explanted and replaced with an ACIOL. Medical history included hypertension. There was no significant family history.

On examination, best-corrected visual acuities were 6/12 in the left eye and 6/36 in the right eye. The IOL with the capsular bag was dislocated in the anterior chamber in the left eye (Figure 1). Intraocular pressures were 16 mmHg in the left and 14 mmHg in the right eye. Axial lengths were 24.13 mm in the left and 24.36 mm in the right eye. Keratometry in the left eye was 8.25 mm and 8.16 mm.

He had some skeletal features suggestive of Marfan's syndrome—height 185 cm, arm span to height ratio of 1.04 (>1.05 in Marfan's) and upper to lower segment ratio of 0.71 (<0.86 in Marfan's). Routine blood investigations, urine analysis, glucose, ECG, and echocardiogram were normal. A cardiologist ruled out systemic and cardiac abnormalities.

He was managed by explantation of the IOL with the bag and insertion of an ACIOL. There was no zonular resistance at the time of surgery. 6 weeks postoperatively, best-corrected visual acuity was 6/4.

Comment

The aetiology and mechanism behind spontaneous luxation of an IOL with the bag, in the absence of any predisposing factors is still uncertain. To date, there have been five such reports.[4–8] The mechanisms in three of these were ocular trauma with high myopia,[4] capsular contraction with pars planitis,[8] and severe rubbing associated with atopic dermatitis.[7] The cases described by Shigeeda *et al*[5] were relatively young with the possibility of a traumatic cataract. The mechanism was thought to be peroperative zonular stress in combination with capsular shrinkage. Nishi *et al*[6] were of the same opinion.

Our patient differs from the previous reports in that he was younger (41 years), the time span was greater (14.5 and 9 years), and he had an anterior dislocation in one and a posterior dislocation in the other eye. In the absence of any known predisposing factors, primary or secondary zonular weakness has been thought to be the likely mechanism.[1,5,6,8] Contraction of the capsular opening occurs in all postoperative patients,[9] and is common with continuous curvilinear capsulorhexsis.[8] Anterior capsular phimosis can lead to an imbalance between the centrifugal and centripetal forces of the capsulorhexsis margin,[3] which may exert continuous centrifugal forces on the zonules causing their rupture.[6] Our patient had an adequate anterior capsule opening. The IOL material and design may also have an effect on capsular shrinkage. Single-piece polymethylmethacrylate (PMMA) lenses are thought to be better than three-piece lenses[5] and acrylic IOLs are associated with less fibrosis

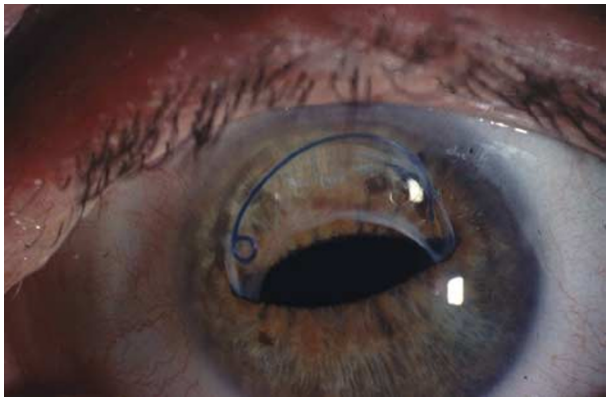

Figure 1 IOL with the capsular bag dislocated in the anterior chamber.

in comparison to PMMA, silicon, and hydrogel lenses.[3] This might be relevant in our patient's left eye, as he had a three-piece lens. However, as previous reports have included single-piece PMMA,[4,6] three-piece PMMA,[5] and acrylic[5] IOLs, no definite conclusion can be drawn. These factors are unlikely to be relevant in his right eye.

The posterior luxation of the PCIOL in our patient's right eye raises the possibility of a bilateral involvement, although it could be a result of previous surgery. He had bilateral developmental cataracts and a retinal detachment in one eye, which are common in Marfan's syndrome and suggest a multifactorial aetiology. Although our patient had no family history, he had some features suggestive of Marfan's syndrome—tall habitus, arm span/height ratio of 1.04, upper/lower segment ratio of 0.71, reduced keratometric power, and increased axial length. Spontaneous luxation of an IOL has not to our knowledge been reported, as a presenting feature in Marfan's syndrome to date. These features are, however, not sufficient to make a diagnosis of Marfan's and it is debatable, whether this is a *forme fruste* of Marfan's syndrome. Mengesha[10] reported ectopia lentis in the absence of a family history and other signs and called it a *forme fruste* of Marfan's, implying that not all clinical features are present and the diagnosis is not secure. As genetic mutation occurs in 15% of cases, a family history may not always be positive. Although the mechanism of IOL-bag luxation in our case is inconclusive, it is most likely a manifestation of Marfan's syndrome.

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Sir,

Limited Wegener's granulomatosis of the orbit: a case study and review of literature

Wegener's granulomatosis (WG) is a severe, noninfectious, granulomatous, and necrotizing vasculitis that classically affects respiratory tracts and kidneys.[1] Clinical manifestations of WG are nonspecific and indistinguishable from a variety of neoplastic, infectious, and inflammatory diseases. The limited form of WG usually lacks renal involvement, frequently involves the head and neck region, and may not always progress to generalized disease with a resultant better prognosis.[2,3] We report an unusual presentation of limited WG of the orbit showing extensive orbital involvement without systemic or sinonasal tract involvement. The histopathology showed a classical triad of vasculitis, necrosis, and granulomatous inflammation with *multinucleated giant cells*, which are rarely seen from the orbital specimen.[4]

Case report

A 56-year-old male presented to us in July 2003 with complaints of right-sided gradually progressive proptosis (since August 2002), associated diminution of vision (since January 2003), and ptosis (since May 2003).