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EC OPHTHALMOLOGY
Case Report

Lens Subluxation in Marfan Syndrome

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Abstract

Marfan disease is a common autosomal dominant connective tissue disorder resulting in mutations of genes required to produce the glycoprotein fibrillin. Several ocular features of Marfan syndrome have been well documented in the literature and indeed it has been determined that fibrillin plays a vital role in stabilising the crystalline lens. This case report details the presentation of a 34-year-old male with progressive visual loss bilaterally over the course of a few months. Examination of the patient revealed a visual acuity reduced to 6/10 bilaterally and ectopia lentis bilaterally. Given the subluxation was severe and occurring superiorly a bilateral phacoemulsification with intra-ocular lens implant procedure with iris suture fixation occurred. This produced successful results. Other common ocular manifestations of Marfan syndrome include retinal detachment and glaucoma. Swift investigation and management of visual disturbances is advised in such circumstances to reduce the risk of any long-term visual deficits..

Keywords: *Marfan Syndrome; Ectopia Lentis; Lens Dislocation*

Introduction

Marfan disease is a common autosomal dominant connective tissue disorder resulting in mutations of genes required to produce the glycoprotein fibrillin.

Case Report

A 34-year-old male with a history of Marfan syndrome was seen in an ophthalmology clinic following referral from the Emergency Department the previous day. The patient reported a progressing blurring of vision in both eyes bilaterally over the past few months with significant deterioration in the past week prompting his attendance to the Emergency Department. He reported also experiencing occasional glare but denied any other symptoms. The patient's past medical history was of Marfan syndrome and no other conditions. The patient was on bisoprolol 2.5 mg for aortic stenosis and mitral valve prolapse secondary to his Marfan syndrome.

On examination visual acuity was reduced bilaterally with 6/10 vision in both eyes. Pupils, occular movements and color vision was normal. IOP were 14 mmg in the right eye and 16 mmg Hg in the left. Examination of the anterior segment with a slit lamp revealed that both lenses were subluxated superiorly. Fundoscopy revealed a normal posterior segment with no retinal abnormalities.

Given that the lens subluxation was severe, it was agreed that the patient was not a good candidate for conservative management with contact lens correction, therefore he underwent a bilateral phacoemulsification with intra-ocular lens implant. The patient's vision returned to normal post-operatively and during the surgery, iris suture fixation of the intra-ocular lens was used to prevent future subluxation.

Discussion and Conclusion

Marfan syndrome is a common autosomal dominant disease which causes weakened connective tissue in a number of organ systems including the eye. It is thought to affect 1 in 5000 people [1]. Specifically, a defect in the in the protein fibrillin 1 (FBN1) results in impaired amalgamation of fibrillin into connective tissue. One of the most common manifestations of Marfan syndrome is ectopia lentis which refers displacement of the eye's crystalline lens, this dislocation can be partial (such as in this case) or complete (termed a luxation). Ectopia lentis occurs in around 60% of patients and is usually bilateral [2]. The displacement can occur in various directions, and can lead to severe complications such as glaucoma, if for example it occurs anteriorly. Symptoms of subluxation intelligibly depend on the direction and severity of dislocation; however they are usually blurred vision, pain and diplopia. Management should be tailored to each individual patient with mild subluxations being managed with refractive correction and surveillance and more severe dislocations being managed surgically, with a number of different specific surgical techniques being used. In addition to ectopia lentis another common ocular complaint of patients with Marfan syndrome are retinal tears or detachments, particularly due to the high myopia patients with Marfan's tend to experience [3]. Therefore, this is something crucial to be wary of when examining these patients.

Learning Points

1. Ectopia lentis is very common occurrence in Marfan syndrome.
2. Ectopia lentis can be managed medically or surgically depending on degree and direction of lens dislocation.
3. Patients with Marfan syndrome should be screened for retinal tears and retinal detachment, given their typical high myopias.

Conflicts of Interest

The author declares no conflicts of interest.

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