Case Report

A Rare Case of Bilateral Microspherophakia

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Abstract

Microspherophakia is a rare bilateral congenital anomaly of the crystalline lens. The condition may be isolated, familial, or it may be associated with systemic affections like Marfan's syndrome, Weil-Marchesani syndrome, hyperlysinemia and congenital rubella. Microspherophakia results in lenticular myopia, lens dislocation, usually inferiorly and inverse glaucoma. We present a case in a 8 year old child who presented with bilateral microspherophakia. Visual acuity in the right eye was counting fingers close to face and in the left eye 6/60.IOP with Perkins applanation tonometer was 30 mmHg in the right eye and 22 mmHg in the left eye, cornea was hazy due to edema, anterior chamber was shallow in both eye patient was managed with emergency lens extraction of the right eye and secondary ACIOL implantation. Left eye was managed by laser peripheral iridotomy. IOP was within normal limits postoperatively in both eyes without any antiglaucoma medications. Postoperatively, best corrected visual acuity in the right eye was 6/18 and 6/9 in the left eye.

Keywords: Microspherophakia, Anterior dislocated lens, Isolated microspherophakia.

Introduction

Microspherophakia is a rare congenital condition of the crystalline lens wherein the anteroposterior diameter is more than the horizontal diameter and the lens assumes a spherical shape.[1] Defective development of the zonules results in their deficiency, increased length, weakness and non-attachment of posterior zonules to the ciliary processes. This could be regarded as a simple arrest of lens development between the fifth and sixth month of intrauterine life.[2,3] This leads to formation of a small spherical lens. Microspherophakia is associated with some systemic affection like Weil-Marchesani syndrome, Marfan syndrome, or isolated microspherophakia.[4]

Case Description

A 8 year old female child was brought to the outpatient department with complaints of diminution of vision in both eyes since her early childhood and complaints of redness and watering from the right eye for 1 week. There was no history of trauma and no history of similar complaints in the past, no history of similar complaints in any siblings and no history of consanguineous marriage. She was full term normal delivery and normal developmental milestones on general physical examination; there were no signs of Marfan's or Weil-Marchesani syndrome. Examination of the right eye revealed mild circumciliary congestion cornea was hazy due to corneal edema. AC was shallow with a dislocated lens and the lens was microspherophakia. IOP in the right eye with a Perkins applanation tonometer was 30 mmHg; fundus glow was seen, but other details were not made out. Examination of the left eye revealed a shallow anterior chamber and microspherophakia. IOP in the left eye with a Perkins applanation tonometer was 22 mmHg; fundus was normal in left eye visual acuity in RE counting fingers close to face and in LE 6/60 best corrected visual acuity in RE counting fingers close to face and in LE 6/36. She was put on I.V mannitol 20% and taken up for lens extraction of the right eye under general anaesthesia, and a secondary ACIOL was implanted after 1 month. Postoperatively, VA in RE was 6/18. Laser iridotomy was done in LE. Postoperatively, IOP was within normal limits without any antiglaucoma medications (Figure 1).

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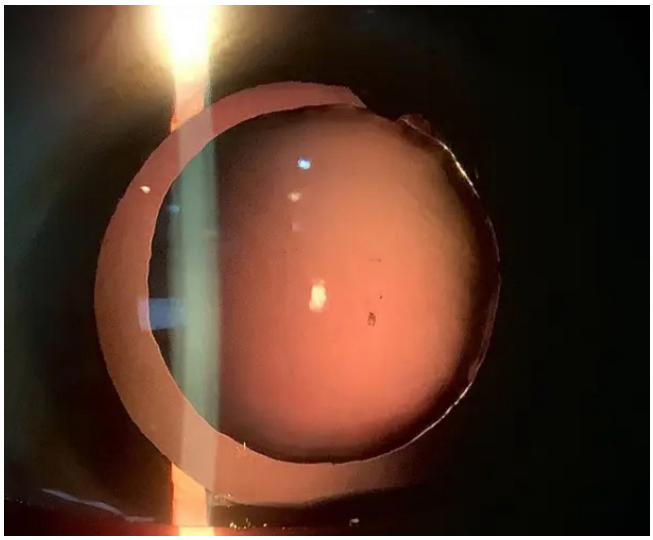

Fig. 1: Microspherophakic lens visible after mydriasis

Discussion

Isolated microspherophakia is a rare condition. It is most often hereditary or familial or integrated into a general malformation syndrome.[5] Investigators have hypothesized that spherophakia occurs when an incompletely developed ciliary body and its loose, elongated zonules do not exert sufficient pressure to flatten the developing lens. The lenses of patients with spherophakia therefore retain a fetal spherical conformation.[2,3] It usually presents in the first or second decade of life with progressive myopia and angle-closure glaucoma due to pupillary block by a microspherophakic lens. Use of miotics causes forward displacement of lens iris diaphragm and causes inverse glaucoma in such patients.[6,7] Familial microspherophakia, generally not associated with other systemic malformations, is inherited as an autosomal recessive trait and associated with ectopia lentis, where the lens is most frequently displaced upwards.8 With our patient, the manifestation of this condition is sporadic or possibly familial, as no previous case was identified in the immediate family.

Conclusion

As there were no other signs or symptoms suggestive of Weil-Marchesani, Marfan, or other syndromes, we concluded that this case is an isolated bilateral ectopic microspherophakia.

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