CASE REPORT

A RARE CASE OF BILATERAL MICROSPHEREPHAKIA

Pandu S[1], Arunkumar B. Desai[2], Sujatha V[3], Srinivas B[4], Muthukumaran R. S[5]

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ABSTRACT: Microspherophakia is rare bilateral congenital anamoly 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 and anterior dislocation of lens of right eye. visual acuity in right eye was counting fingers close to face and in left eye 6/60.IOP with perkins applanation tonometer was 30mmHg in right eye 22mmHg in left eye, cornea was hazy due to edema, anterior chamber was shallow in both eye patient was managed with emergency lens extraction of 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 right was 6/18 and 6/9 in left eye.
KEYWORDS: Microspherophakia, Anterior dislocated lens, Isolated microspherophakia.

INTRODUCTION: Microspherophakia is a rare congenital condition of crystalline lens where in the anteroposterior diameter is more than horizontal diameter and 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, Marfans syndrome or isolated microspherophakia.[4]

CASE DESCRIPTION: A 8 year old female child brought to outpatient department with complaints of diminution of vision in both eyes since her early childhood and complaints of redness watering from right eye since 1week there was no history of trauma and no history of similar complaints in past, no history similar complaints in any siblings and no history of consanguineous marriage. She was full term normal delivery and with normal developmental mile stones on general physical examination there were no signs of Marfan's or Weil Marchesani syndrome. Examination of right eye revealed mild circumciliary congestion cornea was hazy due to corneal edema AC was shallow with dislocated lens and lens was microspherophakia IOP in right eye with Perkins applanation tonometer was 30mmHg fundus glow was seen but other details were not made out. Examination of left eye revealed shallow anterior chamber and microspherophakia, IOP in left eye with Perkins applanation tonometer was 22mmHg 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 right eye under general anaesthesia and secondary ACIOL was implanted after 1 month. Post operatively VA in RE was 6/18.Laser iridotomy was done in LE. Post operatively IOP was within normal limits without any antiglaucoma medications.

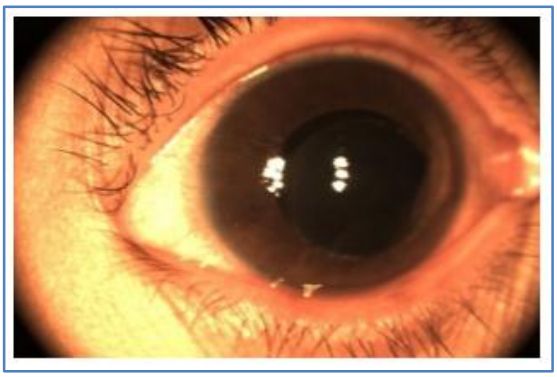

Fig. 1: Dislocated lens in anterior chamber

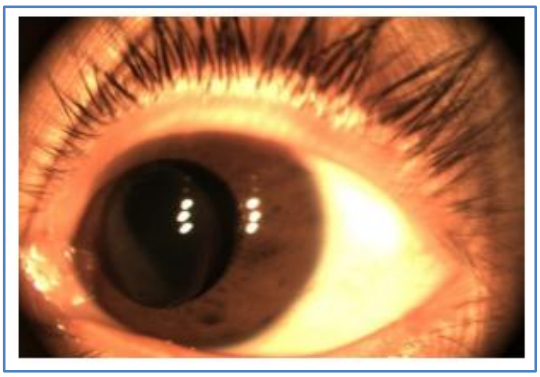

Fig. 2: Microspherophakic lens visible after mydriasis

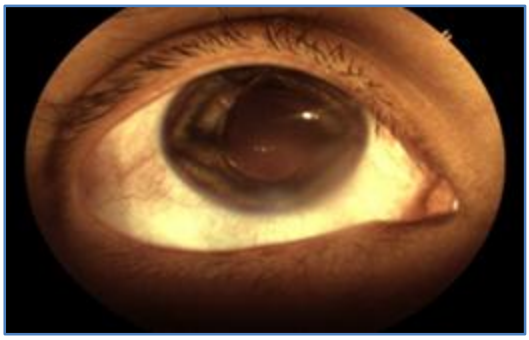

Fig. 3: Post-operative picture showing ACIOL in situ

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 first or second decade of life with progressive myopia, angle closure glaucoma due to pupillary block by microspherophakic lens.Use of miotics causes forward displacement of lens iris diaphragm and cause 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.

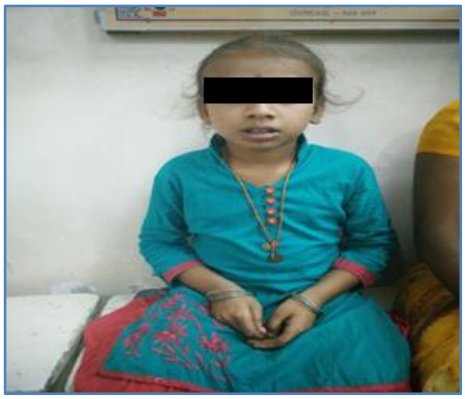

Fig. 4: General physical and systemic examination was normal

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

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AUTHORS:

1. Pandu S.
2. Arunkumar B. Desai
3. Sujatha V.
4. Srinivas B.
5. Muthukumaran R. S.

PARTICULARS OF CONTRIBUTORS:

1. Professor & HOD, Department of Ophthalmology, M. V. J. Medical College of Research Institute.
2. Post Graduate, Department of Ophthalmology, M. V. J. Medical College of Research Institute.
3. Associate Professor, Department of Ophthalmology, M. V. J. Medical College of Research Institute.
4. Assistant Professor, Department of Ophthalmology, M. V. J. Medical College of Research Institute.
5. Post Graduate, Department of Ophthalmology, M. V. J. Medical College of Research Institute.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Arunkumar B. Desai,
Post Graduate,
Department of Ophthalmology,
M. V. J. Medical College & Research Institute,
Bangalore.
E-mail: drarunkumardesai@gmail.com

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