Research Paper Medical Science

Anterior Subluxation of Crystalline Lens in Case of Marfan's Syndrome

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KEYWORDS

INTRODUCTION:

- Marfan syndrome is a pleotropic autosomal dominant genetic disorder that results in weakening of connective tissue in the musculoskeletal, cardiovascular and ocular organ system. They exhibit arachnodactyly , a high-arched palate, and facial abnormalities. The cardiovascular findings range from mild mitral valve prolapse to severe aortic aneurysm or dissection

- The major ocular abnormality in Marfan syndrome is ectopia lentis . The most common direction of dislocation found is superotemporal . Retinal tears and detachments are also quite common in patients with Marfan syndrome.

CASE REPORT:

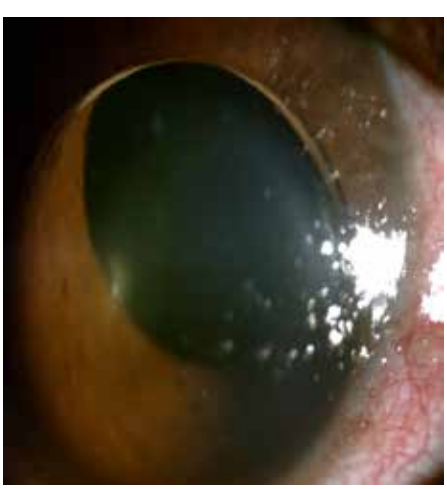
- A 36-year-old male, residing in Kapadvanj , Gujarat presented to our hospital with complaints of pain , redness and dimness of vision in right eye since 2 days.
- Patient was relatively asymptomatic before 2 days and then suddenly developed painful dimness of vision with headache and redness in right eye.
- Patient had h/o left eye pthysical eye since 5 years after development of corneal ulcer abscess.
- There was no history of any trauma or any surgical intervention in right eye.
- No complain of easy fatiguability or any other systemic illness.
- On examination ARMSPAN was relatively higher compared to the height. Patient had high arched palate with macroglossia.
- Ocular movements were free and painless in all directions.
- On ocular examination lid, lacrimal apparatus was normal.
- Conjunctival and circumcorneal congestion was present in right eye. Cornea was clear.
- Normal sized crystalline lens was present in anterior chamber blocking the angle of anterior chamber .

HIGH ARCHED PALATE

OCULAR EXAMINATION:

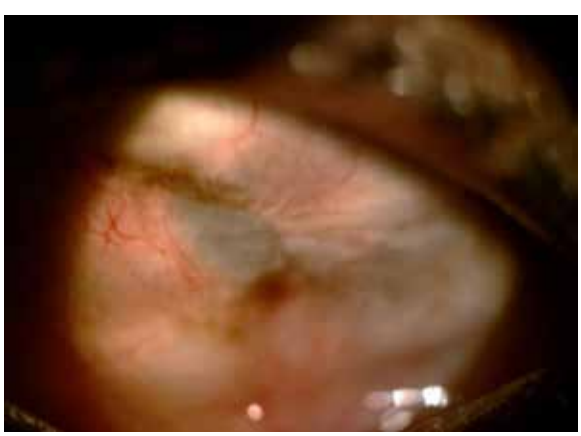
- Pupil was semidilated and fixed with vitreous in pupillary area.
- Fundal glow was visible and it was normal healthy orange.
- Intraocular pressure was raised in right eye which was 54 mm Hg with applanation tonometry.
- Visual acuity in RE was counting finger 2 feet and left eye was PL+ PR faulty.
- Blood pressure was 120/80 mm Hg .

RIGHT EYE



RIGHT EYE

LEFT EYE

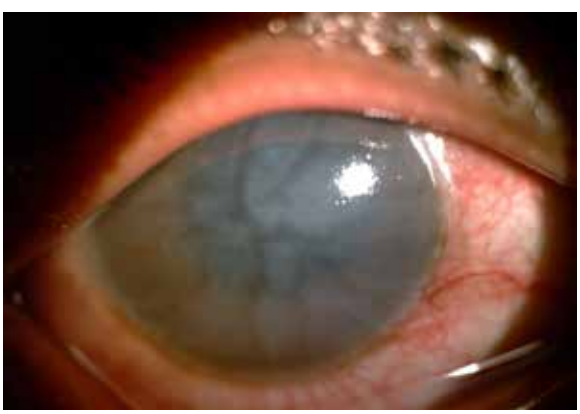


INVESTIGATIONS DONE:

- Blood investigations in the form of complete blood count, plasma blood sugar were normal. X ray chest PA view was within normal limits.
- Hb – 10.0
- TC – 8000
- DC – 68/28/3/1
- RBS- 101
- B. Urea – 26
- S. Creatinine – 0.8
- S.bilirubin – 1.2
- S. HIV – NR
- S.HBsAg – NR
- ECG showed LVH strain pattern with broad QRS complex

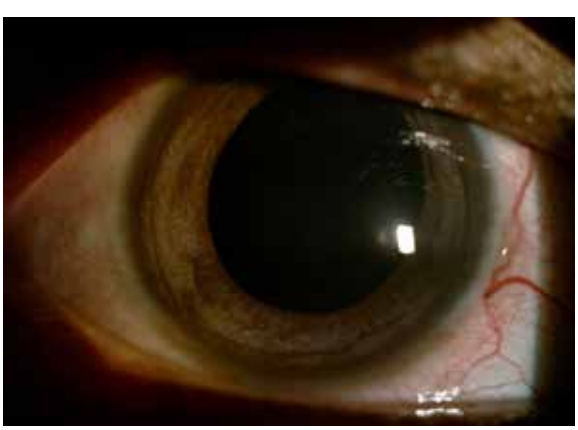
TREATMENT GIVEN:

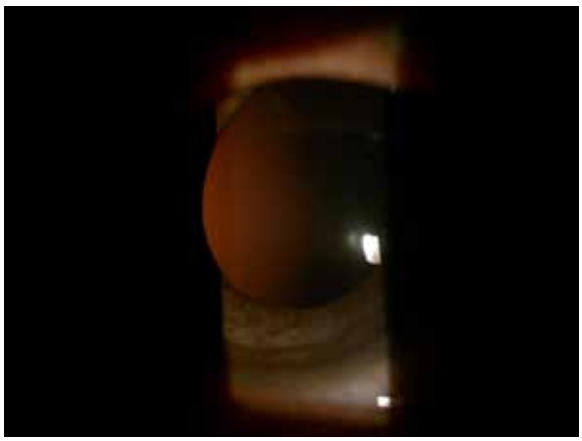
- Patient was given injection intravenous mannitol 350 ml stat over 20 mins.
- After injecting mannitol , pressure decreased to 35 mm Hg in right eye.
- Then patient was operated for lensectomy by making sclerocorneal tunnel with anterior vitrectomy without intraocular lens insertion and kept aphakic.
- Patient had corneal oedema on post operative day 1.
- Patient was better postoperatively with clear cornea and vision improved to 6/24 on snellen's chart with +12.0 D sph correction after 3 weeks postoperatively.

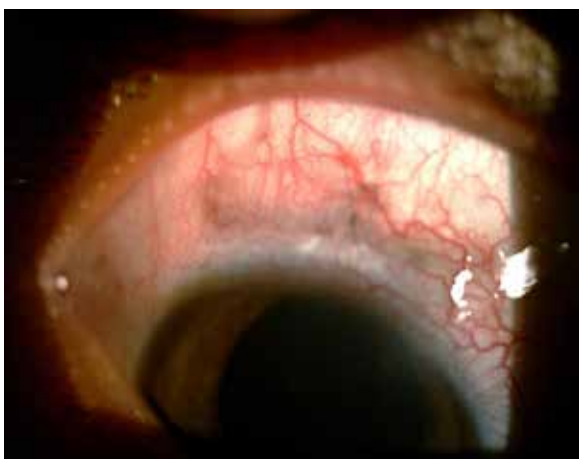


POST-OP DAY -1

3RD WEEK POST-OP







DISCUSSION:

- The abnormality in over 80% of patients with Marfans disease involves defects in the protein fibrillin 1 (FBN1) on chromosome 15, a structural component of microfibrils found in connective tissue throughout the body.
- The systemic manifestations in patients of Marfan syndrome are that they are tall with long, flexible extremities and marked scoliosis. They exhibit arachnodactyly (spider fingers) with the ability to dramatically encircle the wrist (Walker-Murdoch sign). In addition, they often have pectus excavatum, a high-arched palate, and facial abnormalities. The cardiovascular findings range from mild mitral valve prolapse to severe aortic aneurysm or aortic dissection; and severe cardiovascular complications are the primary causes of mortality in Marfans syndrome.
- In eye , Marfan patients exhibit abnormal cilliary processes with absent or severely disorganized zonules. This pathology was found to be positively correlated with lens subluxation. The most common direction of dislocation on examination is superotemporal. In addition, there may be secondary complications from lens movement such as phacolytic uveitis from posterior subluxation of the lens to the vitreous.With regard to ectopia lentis, a number of treatment options exist. Mild subluxation allows for near normal vision with the patient seeing through the phakic portion of the pupil. Surgery is indicated when the lens position causes irregular astigmatism and glare, when the lens is posteriorly dislocated into the vitreous, when the lens is dislocated anteriorly and causes secondary glaucoma.

CONCLUSION:

Thus from this study it can be proved that by doing early intervention in case of Marfan's syndrome by doing vitrectomy and lensectomy we can prevent complications like secondary glaucoma and get a better final visual outcome.

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