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Bilateral Opposite Lens Dislocation in Marfan Syndrome : A Case Report

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Abstract:- One of the main diagnostic criteria for Marfan syndrome is ectopia lentis , which can range from a moderate displacement without symptoms to a substantial dislocation where the lens's equator is in the pupillary axis. This work's goal is to present the case of a 15-year-old patient diagnosed with Marfan syndrome who presented with bilateral opposite lens dislocation.

Keywords:- *Ocular Hypertonia, Bilateral Lens Dislocation, Marfan Syndrome, Zonular Instability.*

I. INTRODUCTION

The first case of a kid with Marfan Syndrome was reported by Antoine-Bernard-Jean Marfan in 1896 [1]. This is an autosomal dominant disorder with musculoskeletal, ophthalmological, and cardiac symptoms. It manifests as a result of mutations in the FBN1 gene, which codes for the protein fibrillin, which is essential to the connective tissue's ultrastructure [2, 3]. Fibrillin gives the connective tissues of the eyes flexibility and force-bearing structural stability.

Ectopia lentis has been identified as a significant diagnostic criteria for Marfan syndrome, and it is present in roughly 60–68% of cases. It can be used to diagnose the condition or rule it out in as many as 86% of cases [4, 5].

Ectopia lentis is often bilateral, symmetric, and non-progressive [6]. It can range from a little, asymptomatic displacement only observed in post-pupillary dilation to a major dislocation where the lens's equator is positioned in the pupillary axis, resulting in monocular diplopia. Although the dislocation is typically superotemporal, it can also occur posteriorly and result in vitreous traction on the retina, which can induce chronic vitreitis and chorioretinal inflammation. Forward dislocation of the lens into the pupil or the anterior chamber can cause pupillary block, which can lead to the development of acute or chronic closed angle glaucoma [7, 8].

II. CASE REPORT

A 15-year-old male arrived at our institute's emergency room complaining of unilateral redness, photophobia, ocular pain of the right eye. and decreased visual acuity on both eyes.

He had been diagnosed in childhood with cardiovascular symptoms, such as aortic root dilatation and mitral valve prolapse related to Marfan syndrome.

His ophthalmologic history involves the usage of hard contact lenses to treat her high myopia, which is -10.00 D in the right eye and -9.00 D in the left eye. The interrogation does not reveal any notion of trauma

On ophthalmologic examination the visual acuity of his right eye was hand motion on the right eye and counting fingers on the left eye. The Intraocular pressure measured at that moment was 40 mmg on the right eye and 12 mmHg on the left eye.

On slit lamp examination at the right eye, we find conjunctival hyperemia with complete dislocation of the lens in the anterior chamber and whose equator is visible over 360° in front of the iris, corneal edema, and areflexic mydriasis [Figure 1].

Examination of the left eye reveals a non-inflamed eye, a clear cornea, no Tyndall in the anterior chamber, with lower dislocation of the lens in the posterior chamber, zonules clearly visible superiorly [Figure 2].

On fundus examination of both eyes there was an oblique insertion of the optic disc, and one myopic crescent surrounding the optic nerve, posterior staphyloma. There were also macular pigmentary abnormalities with a visible choroidal fundus in the posterior pole.

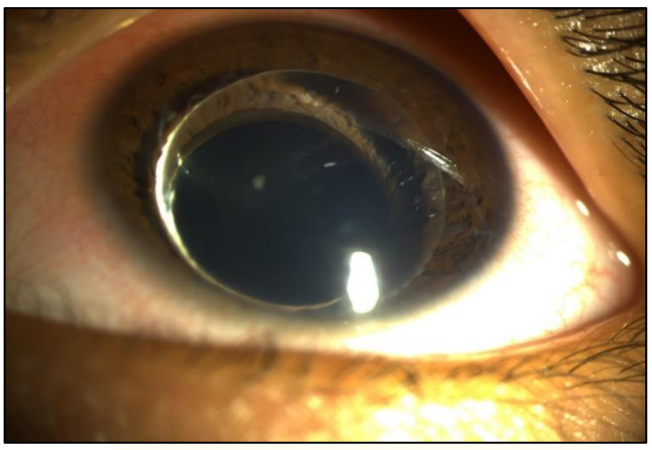

Fig 1 In this Figure of the Right Eye we can see the Dislocation of the Lens on the Anterior Chamber

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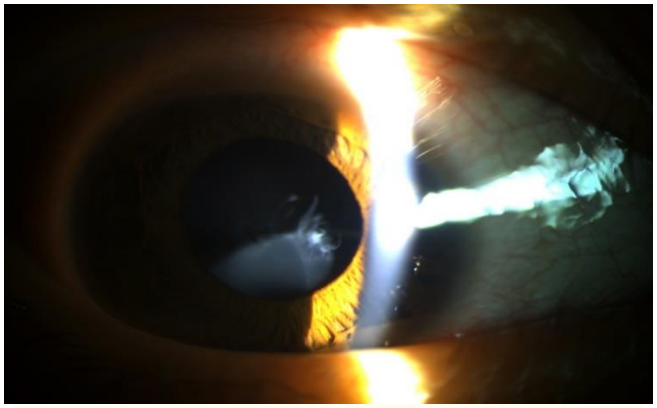

Fig 2 In this Figure of the Left Eye, we can see the Inferior Dislocation of the Lens on the Posterior Chamber

The gonioscopic examination of the right eye found a closed angle over 360 degrees, on the other hand it was open at the level of the left eye.

Morphologically, the patient presented a marfanoid appearance with an exaggeration of the length of the limbs in relation to the trunk [Figure 3], the wirst sign was positive [Figure 4]. He presented also flat foot valgus with arachnodactyly [Figure 5].

Fig 3 Marfanoid Appearance with an Exaggeration of the Length of the Limbs in Relation to the Trunk

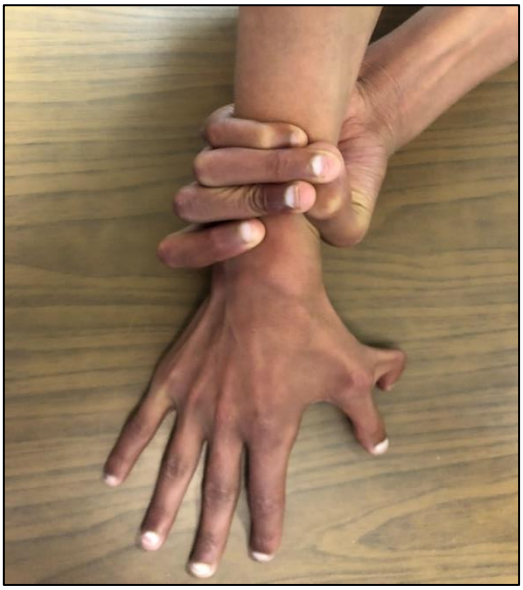

Fig 4 Wirst Sign Positive

Fig 5 Arachnodactyly Appearance on the Left Hand

The patient was diagnosed with high myopia with bilateral anterior and posterior dislocation of the lens causing decreased bilateral visual acuity and an acute crisis by closure of the angle in the right eye.

Medical treatment was instituted urgently combining anti-edematous eye drops, local hypotonizing agents (beta blockers) and general hypotony (mannitol), miotic eye drops in the right eye to prevent the lens from passing into the posterior chamber, the patient was then urgently admitted to the operating room for lensectomy associated with anterior vitrectomy. The left eye was scheduled for endoocular surgery.

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The post-operative course was marked by a normalization of intraocular pressure, measured at 16 mmHg on both eyes, regression of corneal edema. Correction aphakia glasses have been prescribed later.

III. DISCUSSION

Marfan syndrome is a hereditary connective tissue disease transmitted autosomal dominantly, with incomplete penetrance and variable expressivity.

Its prevalence varies from 1/5000 to 1/20000[9].

It manifests itself in the organs derived from the mesoderm, mainly the eye, the skeleton and the aorta.

The first clinical signs most often observed in children were high-arched palate (82%), arachnodactyly (71%) and flat feet (59%).

The demonstration of significant dilatation of the aorta (47%) or ectopy of the lens (35%) could also be a way of discovering the disease.

The ophthalmological criteria for diagnosing disease Marfan have a major criterion and minor criteria [9].

The major criterion is lens ectopia, which has a great diagnostic value: it is present in 50 to 80 % of patients affected. Minor criteria are a cornea abnormally flat, the increase in the axial length of the eyeball and hypoplasia of the iris or ciliary muscle.

Anterior lens dislocation can occur spontaneously or after even minimal trauma. It constitutes a therapeutic emergency because of the risks of glaucomatous optic neuropathy secondary to hypertonia acute and secondary alterations of the corneal endothelium lens contact and ocular hypertonia. It can also occur posteriorly and result in vitreous traction on the retina, which can induce chronic vitreitis and chorioretinal inflammation.

IV. CONCLUSION

As ectopia lentis may be the first sign of a more serious systemic disease like Marfan syndrom. It is important to determine the etiology for appropriate patient management. Lens dislocation in Marfan syndrome is usually bilateral and occurs most often in the superotemporal direction, though other directions are not uncommon. Only 20% will dislocate in a non-superior direction [10]. Additionally, ectopic lentis can cause major complications within the eye, including significant refractive shifts, pupillary block glaucoma, retinal damage, and blindness.

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