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Bilateral Nasal Lens Sub-luxation in Marfan Syndrome: A Rare Case Report

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Abstract: *Marfan syndrome (MFS) is a heritable genetic disorder of connective tissue which causes spectrum of disorders involving musculoskeletal, cardiac, and ocular system predominantly. Hereby We report a case of an eleven year old boy who presented to Sankara Eye Hospital Krishnankovil , Virudhu Nagar District , Tamil Nadu State, in October 2018, with complaints of poor vision of 2 years duration both eyes. On Examination of patient features revealed which were similar to Marfan syndrome and bilateral intra ocular lens subluxation . The patient was refracted and glasses recommended which improved his vision were prescribed. The patient was referred to Cardiology , Orthopaedic , Dental departments respectively for multidisciplinary approach to prevent complications and further management.*

Key Words: *Marfan syndrome, Ectopia lentis, Ocular manifestation*

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I. Introduction

Marfan syndrome (MFS) is a genetic disorder of the connective tissue. Marfan syndrome is an autosomal dominant systemic disorder of connective tissue. Children affected by the Marfan syndrome carry a mutation in one of their two copies of the gene that encodes the connective tissue protein fibrillin-1 (FBN 1)[1]. Fibrillin, a glycoprotein provides force bearing structural support and elasticity of the ocular connective tissues .

It mostly affects skeleton, lungs, eyes, heart, and the aorta. Affected individuals often are tall and slender, have arachnodactyly, scoliosis, and either a pectus excavatum, pectus carinatum, or ectopia lentis in eyes[2]. Marfan syndrome was first described by Antonie-Bernard Marfan in an 1896 case report of a young girl with unusual musculoskeletalfeatures[3].

The ophthalmic features of Marfan syndrome were first described in 1914 by Boerger a Paediatrician. Worldwide, the incidence of Marfan syndrome is approximately 7-17/100,000[4.] In view of importance of the ocular manifestations of Marfan syndrome, we decided to report the case of an eleven year old boy from Krishnankovil who presented with bilateral lens subluxation.

II. Case Report

A 11 year old boy was brought by his mother to Sankara Eye Hospital Krishnankovil on October 10, 2018 with complain of poor vision in both eyes after noticing that he watched television and objects close to his eyes with 2 years duration. The patient had no history of pain, fatiguability , breathlessness ,trauma, no past medical history . His intelligence normal and behavior is hyperactive .He had no previous ophthalmic examination. He is first and last child. Child family history was negative for Marfan syndrome and lens subluxation. He is product of normal spontaneous vertex delivery.

On general Examination , the patient was tall and thin stature. Long extremities showing spidery fingers (arachnodactyly) , long toes with prominent finger joints , elongated face ,high arched palate . The height of the patient was 153 cm while the arms length was 155 cm.

The examination of CNS intact , Chest was clear and no abnormality detected in abdomen . On examination of cardiovascular system revealed left parasternal heave, both S1 and S2 sounds are audible however S2 was louder.

Patient underwent detailed ocular examination in the hospital which revealed visual acuity of 5/60 improved to 6/18 with pin hole in both eyes , further no improvement in visual acuity after refraction. The anterior segment was quiet normal in both eyes ,lens was clear ,pupils briskly reactive. Further examination post dilatation of both pupils revealed nasal subluxation of clear crystalline lens in both eyes. The zonules stretched and remain intact. Fundus examination after full mydriasis revealed no abnormality in the retina and pink optic disc, cup disc ratio was 0.3:1 in both eyes.

A subjective refraction and cycloplegic examination was done patient accepted -2.50 sph

-2.00x10^{o}cyl with improvement to 6/18 in both eyes. The Recommended glasses prescribed to patient . Patient was advised for a regular ocular examination every three to six months. In view of findings in cardiovascular examination patient was advised to cardiologist in view of further management.

Figure 1: Full view of the patient

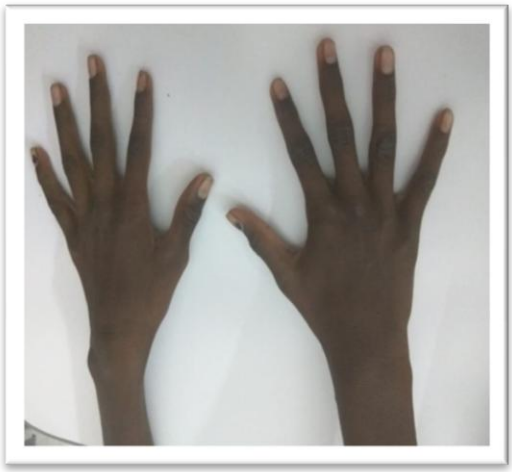

Figure 2: Arachnodactyly (Long Fingers).

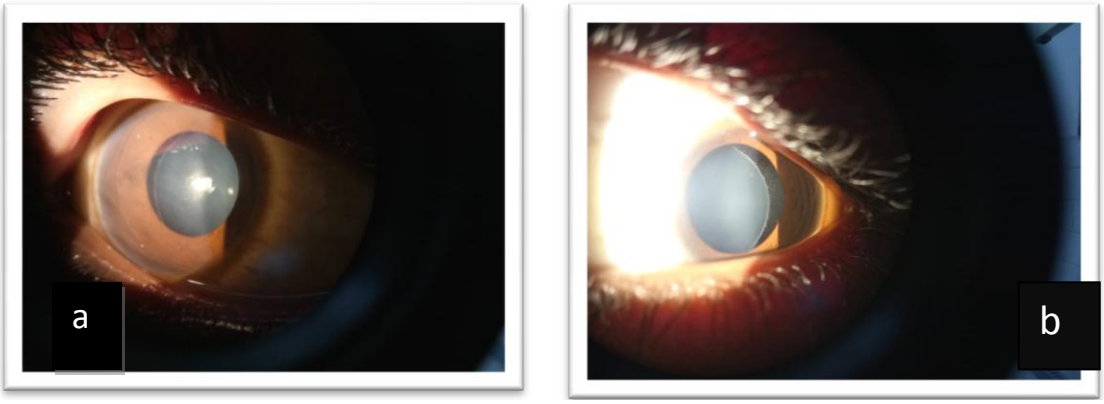

Figure 3: (a) Nasal subluxation in Right Eye
(b) Nasal subluxation in Left Eye

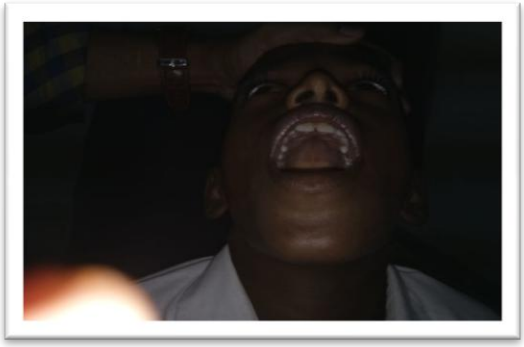

Figure 4: High arched palate

III. Discussion

Ectopia lentis is a displacement or malposition of crystalline lens of the eye in normal position. A partial displacement of lens is called subluxation while a complete displacement is called dislocation .Visual acuity in ectopia lentis patients varies with the degree of malposition of lens.

The presence of Ectopia lentis is a major criterion for the diagnosis of Marfan syndrome according to Ghent criteria, Which eventually establishes diagnostic criterion for Marfan's syndrome in 86% of cases[5], Which was present in our case.

Marfan syndrome is the most frequent cause for inherited ectopia lentis[6.] Ectopia lentis in Marfan syndrome patients usually bilateral, symmetric, supero temporal in location, and non progressive entity in 50-

80%of patients[7]. It varies from mild asymptomatic displacement to significant subluxation resulting in monocular diplopia[8].

Common non surgical interventions includes refractive correction and application of miotic drugs. Surgical indication for lens extraction includes lens opacity with poor visual function, anisometropia, impending total luxation of lens, non- correctable refractive error and lens- induced glaucoma or uveitis.

In the case highlighted above, the presenting complaint of poor vision of patient was related to bilateral lens subluxation which was bilateral nasal subluxation in our case which is rare presentation in Marfan syndrome. In case of Marfan syndrome, it is difficult to detect displacement of crystalline lens prior to poor papillary dilatation even after instillation of atropine eye drop[9]. Even with the lens of normal transparency there is often amblyopia and it is caused by optical changes due to fact that equator of the lens occupies the optical centre of the lens.

Our patient did not show any other ocular finding except bilateral nasal lens subluxation on examination, So we decided to temporize on the issue of lens subluxation in view of fact that patient's vision improved with recommended glasses and also lens transparency clear in both eyes.

The patient was advised to follow up with an ophthalmologist for early detection of ocular features. The need to prevent the ocular complication is the justification for our plan to review the patient periodically . Follow up with cardiologist and orthopedician was suggested to improve quality of life and to detect life threatening complications like dissection of the aorta early. The family members were also examined to rule out the cases of Marfan syndrome.

IV. Conclusion

Ophthalmologist has a very crucial role to play both in diagnosis as well as treatment of Marfan syndrome. There is need to effectively manage ocular features of Marfan syndrome so as to prevent amblyopia and preserving the vision .

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