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Case Report

The Mystery of Ectopia Lentis -A Rare Case Report

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Abstract

We report a case of a 12-year-old male with hematological, ocular, dermatological, dental and gastrointestinal involvement. To the best of our knowledge and after an extensive internet search, this probably is the first case of subluxation of the natural crystalline lens (NCL) with so many systemic associations described plus the pattern of subluxation itself.

Keywords: Ocular; Systemic; Associations

Case Report

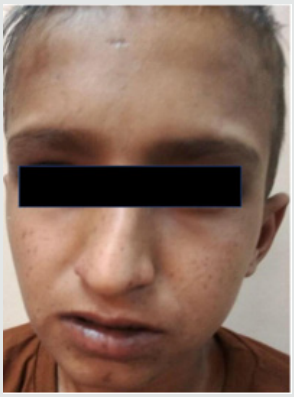

Figure 1

A 12-year-old male (Figure 1) receiving treatment for gastroenteritis from the department of pediatrics (at our institute) presented to us with a history of itching bilateral eyes since his admission to the hospital. Secondly, the department of pediatrics diagnosed the child to be suffering from aplastic anemia and was referring him to a higher center for further management. On the contrary, old records bought by the patient's father and mother showed that the patient was diagnosed as a case of thalassemia from some other institute and was receiving treatment in the form of regular blood transfusion from that institute. Further, the patient had multiple attacks of diarrhoea in the past few years for which he was admitted in hospital many times. The diarrhoea was not related to any particular meal. He was also receiving treatment from dental department for grade 3 mobility of lower anterior teeth. The positive findings on his general physical and systemic examination were thin sparse hairs on the scalp (Figure 2), facial freckles (Figure 3), and barrel shaped chest (Figure 4). His blood sugar, renal function tests, liver function tests, X ray chest, electrocardiography, vitamin B12 plus folate and zinc levels, C reactive proteins and serum electrolytes which were already carried out were within normal limits. The patient's parents were farmers by occupation and did not give a history of themselves suffering from any major illness in the past. The patient's family history revealed that one of the siblings had died at a younger age because of some illness, though no detail or hospital records of the disease were available with the parents. There was no other significant medical, surgical, traumatic or drug usage history.

Figure 2: Thin sparse hairs on the scalp.

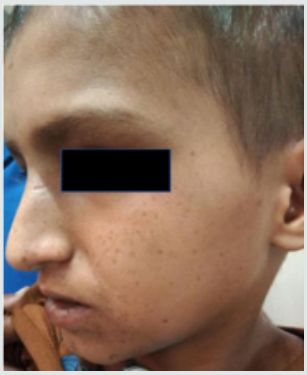

Figure 3: Facial freckles.

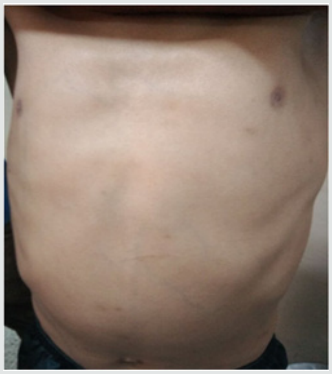

Figure 4: Barrel shaped chest.

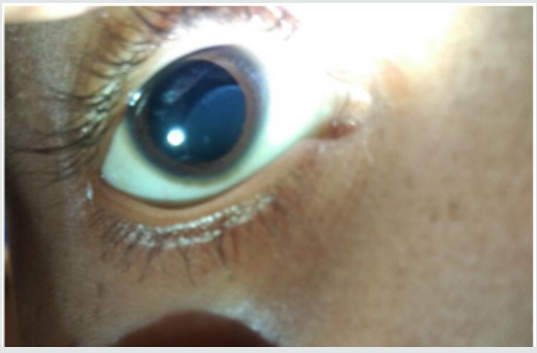

Figure 5: Inferonasal subluxation of the NCL.

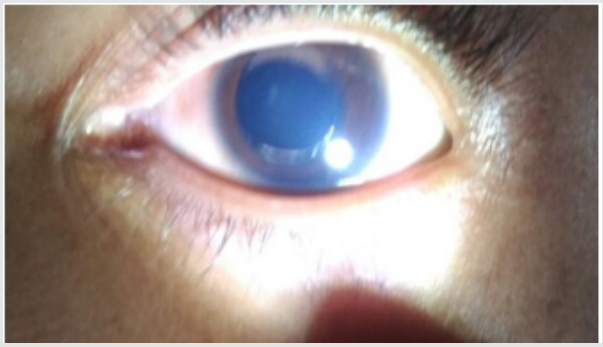

Figure 6: Anterior subluxation of the NCL.

Ocular examination was carried out. His visual acquity was finger counting at 2 meters bilaterally with no improvement on pinhole and refraction. Slit lamp examination of the right eye (Figure 5) revealed inferonasal subluxation of the NCL with a shallow anterior chamber while the left eye (Figure 6) had subluxation of the NCL into the anterior chamber with a shallow anterior chamber. The intraocular pressure and axial lengths of both the eyes were within normal limits. The patient did allow us to do gonioscopy. Bilateral fundus could not be seen because of media haze, though the NCL bilaterally were not cataractous nor was there any intraocular inflammation. Bilaterally, the ocular movements and pupillary reaction were normal. The itching in his eyes was as a result of allergic conjunctivitis for which we prescribed him antihistaminic eye drops. Clinical features and investigations for diagnosis of homocysteinuria, marfans syndrome and ehler danlos syndrome did not reveal any abnormality. Facility for ultrasound biomicroscopy (UBM), Optical coherence tomography (OCT), B scan ultrasonography and genetic studies were not available at our institute. We could have planned for his NCL removal and a probable intraocular lens implantation under guarded visual prognosis, but the patient wanted all his treatment modalities to be done in single institution, and his referral center had all the facilities.

Discussion

Ectopia lentis is displacement of the lens from its normal position. Ocular causes of lens subluxation include trauma, high myopia, buphthalmos, anterior uveal tumors, pseudoexfoliation syndrome, and hypermature cataracts [1]. Systemic disorders associated with ectopia lentis include Marfan syndrome, homocystinuria, Weill-Marchesani syndrome, and Ehlers-Danlos syndrome [2].

Spontaneous dislocation of the NCL is an extremely rare entity, especially its dislocation to the anterior chamber. This dislocation to the anterior chamber can lead to various complications like corneal edema, pupillary block glaucoma and anterior uveitis which requires urgent removal of the NCL [3]. Surgical procedures such as intracapsular cataract extraction, limbal or pars plana lensectomy and anterior vitrectomy and suturing of the haptics to the sclera have been the surgical modalities in use. Femtosecond laser assisted cataract surgery, glued intraocular lenses, instead of conventional scleral fixation of IOLs with sutures for fixation are another useful advancement in treatment [4].

Conclusion

The presence of ectopia lentis with multisystem involvement in our patient could be a part of a syndrome complex or may be an isolated entity it still remains a mystery for us. Ectopia lentis with hematological involvement has probably not been reported before (to the best of our knowledge).

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