Ectopia Lentis in Glucose-6-Phosphate Dehydrogenase (G6PD) Deficiency

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Glucose-6-phosphate dehydrogenase (G6PD) deficiency is an X-linked disorder associated with haemolytic anaemia of varying severity due to defect in an enzyme glucose-6-phosphate dehydrogenase[1]. The various ocular manifestations reported in literature in G6PD deficiency include lenticular opacities[2,3], congenital colour blindness[4], yellowing of the conjunctiva, predisposition to the development of proliferative diabetic retinopathy[5], and development of pterygium[6]. We hereby report a case of ectopia lentis in a patient with G6PD deficiency. To the best of our knowledge this association has not been reported before.

CASE PRESENTATION

A 4 year old male patient presented with diminution of vision in both eyes noticed recently by parents. The child had milestones at the normal age. There was no history of mental retardation, early deaths or similar complaints in other family members and there was no history of consanguinity. The best corrected visual acuity was 6/36 in the right eye (with +14D spherical and +1D cylinder at 90 degrees) and 6/24 in left eye (+14 D spherical lens). Color vision was normal with Ishihara charts.

The patient had deep anterior chamber in both the eyes with bilateral infero nasal sub-luxation of the clear crystalline lens (Figure 1,2). The lens margins were bisecting the pupil in both the eyes. The zonules were missing from 6 to 1'o clock in the right eye and 8 to 4'o clock in the left eye. There were no signs of intra-ocular inflammation. The iris in both the eyes were normal in architecture with no transillumination defects. The pupils were normally shaped and had brisk reaction to light and near response. The fundus examination was unremarkable in both the eyes.

There were no systemic complaints but the patient was a known case of G6PD deficiency. A thorough pediatric evaluation was done to rule out systemic abnormalities. There were no skeletal abnormalities and child showed normal developmental milestones. The echocardiography was unremarkable. The child had hemoglobin of 13.4 gm/dL and a normal morphology and count on peripheral smear examination. The serum levels of Homocysteine were with in normal range and urine examination was negative for amino acids. The patient was diagnosed as having ectopia lentis with ametropic amblyopia. The patient underwent bilateral pars plana lensectomy with anterior vitrectomy under general anaesthesia after informed consent. The patient was subsequently prescribed aphakic glasses and referred for amblyopia therapy.

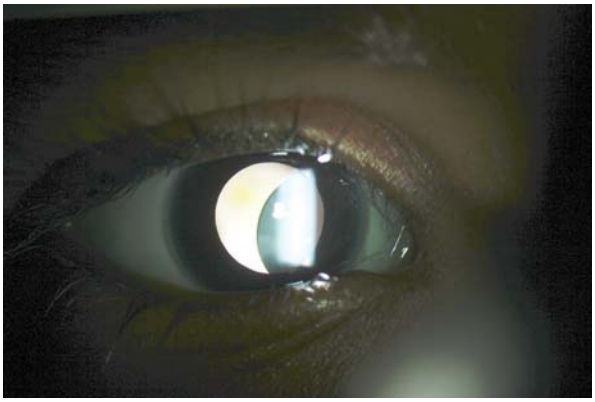

Figure 1: *Photograph of right eye showing subluxation of clear crystalline lens extending from 6 to 1'o clock hours. The subluxated lens is bisecting the pupil.*

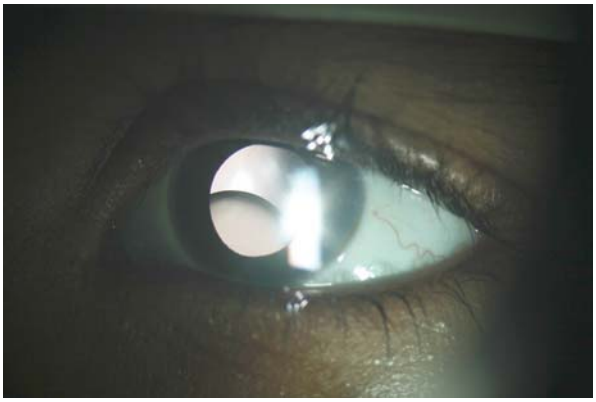

Figure 2: *Photograph of the left eye showing infer-nasal subluxation of clear lens from 8 to 4 'o clock and absent zones.*

DISCUSSION

Ectopia lentis is not an uncommon entity especially in referral practice and can have varied etiologies. The ectopia lentis can be due to genetic, systemic or ocular disorders[7].

Various ocular disorders have been described as the cause of ectopia lentis, most common being ocular trauma. Other common ocular conditions associated with ectopia lentis include retinitis pigmentosa[8], aniridia[9], dominantly inherited blepharoptosis and high myopia[10] and persistent

pupillary membrane[11]. The normal ocular examination and absence of any other signs of trauma ruled out the ocular causes of ectopia lentis in our case.

The common systemic conditions associated with ectopia lentis include Marfan's syndrome, Homocystinuria, Weil Marchesani syndrome, hyperlysinemia and sulfite oxidase deficiency. No systemic abnormality was detected in this child on thorough paediatric examination.

In the absence of ocular and systemic causes we suspected the ectopia lentis to be of genetic origin. The central normal sized pupil excluded ectopia lentis et pupillae in our case. The other genetic condition associated with ectopia lentis is Simple ectopia lentis, which has bilateral symmetric supero-temporal displacement of the lens. The infero-nasal subluxation in our case ruled out the Simple ectopia lentis.

Though it is difficult to prove the direct causation between ectopia lentis and G6PD deficiency, the exclusion of all the known causes of ectopia lentis led us to believe that the ectopia lentis could be associated with G6PD deficiency. Whether there is actual correlation between the two entities or mere coincidence remains to be determined.

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Academic Calendar of Meeting & Conferences 2018 - 19

May 5th, 2018	DOS Evening CME – Hotel Royal Plaza, C.P
July 15th, 2018	1st DOS Monthly Meeting, Dr. R.P. Centre for Ophthalmic Sciences
19th August, 2018	2nd DOS Monthly Meeting & AGBM, Army Hospital (R&R)
September 15th, 2018	**DOS Evening CME-Hotel Crowne Plaza, Rohini**
September 30th, 2018	**3rd DOS Monthly Meeting, DDU Hospital**
October 28th, 2018	**4th DOS Monthly Meeting, Sir Ganga Ram Hospital**
17-18th November, 2018	**DOS Winter Conference**
November 25th, 2018	**5th DOS Monthly Meeting, Venu Eye Institute & Research Centre**
December 30th, 2018	**6th DOS Monthly Meeting, Centre For Sight**
January 27th, 2019	**7th DOS Monthly Meeting, Bharti Eye Hospital**
February 24th, 2019	**8th DOS Monthly Meeting, Guru Nanak Eye Centre**
March 31st, 2019	**9th DOS Monthly Meeting, Dr. Shroff's Charity Eye Hospital**