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Dr. Anukriti
PGT 2ND Year, MGMMC, Kishanganj, Bihar, India

Prof. Dr. RN Saha
Professor and HOD
Dept. of Ophthalmology, MGMMC, Kishanganj, Bihar, India

Corresponding Author:
Dr. Anukriti
PGT 2ND Year, MGMMC, Kishanganj, Bihar, India

Case report on delayed presentation of primary congenital glaucoma

Dr. Anukriti and Prof. Dr. RN Saha

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Abstract
An asymptomatic 5 year old child presented to OPD with corneal haze, megalocornea and raised intraocular pressure. On fundus examination there was bilateral advanced optic disc damage. Surgical interventions done included Trabeculotomy and Trabeculectomy in the left eye and Trabeculectomy alone was done in the right eye. As in this case asymptomatic nature of the disease led to it's late presentation which itself is a cause for advanced disc damage. Therefore awareness towards the subclinical subtle signs of congenital glaucoma makes it a point of concern for pediatricians to do proper screening and referral.

Keywords: primary congenital glaucoma, delayed presentation, optic disc damage

Introduction
Primary congenital glaucoma is a rare disorder which affects infants and younger children. Especially seen predominantly in population practicing inbreeding and consanguinity. In India it's presentation is most of the times severe with severe corneal edema.

Case Report
A 5 year old male patient from outskirts of Bihar, was brought by his parents to Eye OPD of our Tertiary care centre with complaints of an increase in the size and gradual discolouration of both the eyes. The child was having no signs of discomfort and was completely asymptomatic. On taking birth history it was found that he was born out of a non-consanguineous marriage, full term normal vaginal delivery, normal birth weight and milestone. No surgical or medical history. Vision couldn't be elicited but on retinoscopy the child was found to be Myopic. On examination the child was conscious, co-operative and non-irritable. No signs of Photophobia or blepharospasm was present. Extraocular movements were full and was no squint in any eye. On Torchlight examination the cornea was found to be clear and anterior segment structures were fairly visible. On slit lamp examination megalocornea was present with mild corneal haze in both the eyes and there were areas of bluish discolouration of sclera due to focal thinning.
Pupillary examination revealed a Relative Aferent Pupillary Defect in left eye. Applanation Tonometry revealed an Intra Ocular Pressure of 52mmHg in right eye and 60mmHg in left eye. Central Corneal thickness were found to be 630microns and 590microns in right and left eye respectively. Zeiss three mirror gonioscopic examination was found to be normal. Dilated Fundoscopic examination revealed Vertical cup disc ratio of 0.7 and 0.8 in right and left eye respectively. Axial length measured was 26.05mm in right eye and 27.62mm in left eye. Diagnosis was made of Late presentation of primary congenital glaucoma. Topical Antiglaucoma drugs (0.5% Timolol and2% Dorzolamide) were started and he was posted for Combined Trabeculotomy and Trabeculectomy in left eye. Corneal diameters noted preoperatively for right eye was 15.5mm and 15mm vertical and horizontal respectively , and for left eye were 16mm and 16mm vertical and horizontal respectively. The surgery went uneventful. Following this the right eye was also posted for Trabeculectomy alone. Right now the child is fixating fairly and following light bilaterally with IOP of 18mmHg in right eye and 16 mm Hg in left eye.

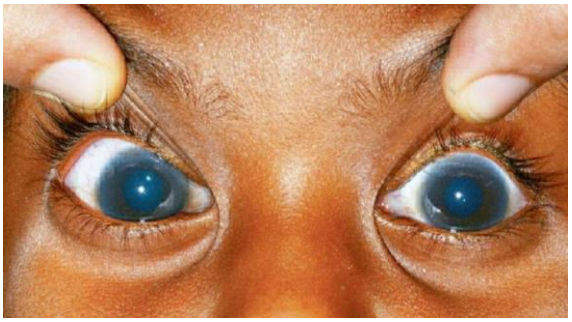

Fig 1: Corneas clear with visible anterior segment structures. Pupil here is pharmacologically dilated

Discussion

We are reporting this case as it is one of the rare presentations of Primary congenital glaucoma with fairly no signs and symptoms except globe enlargement. Minimal and subtle signs of the disease led the parents to ignore it and consequently led to the presentation of the disease at such an advanced stage. If there would have been proper awareness and pediatric screening facilities then the consequences could have been avoided. Compared to other pediatric glaucomas, primary congenital glaucomas have a good visual prognosis with adequate and timely treatment. It is of utmost priority to diagnose congenital glaucomas early and provide proper treatment. High IOP for a long time may lead to irreversible optic nerve damage, corneal opacity, staphyloma, refractive errors and amblyopia. In this case early diagnosis was not possible due to the asymptomatic nature of the disease and it's late presentation.

- The close differential diagnoses include Juvenile open angle glaucoma and Megalocornea.
- Juvenile open angle glaucoma occurs in age group of 5to35yrs. In Juvenile open angle glaucoma there is no enlargement of globe or cornea.
- Another differential diagnoses include Megalocornea which is an inherited disorder with bilaterally symmetrically enlarged but clear corneas. Children presenting with Megalocornea have IOP within normal range and have a fairly clear cornea.

Combined Trabeculectomy and Trabeculotomy was advised due to High IOP and a large cup disc ratio. This Combined surgery was found to have very high success rates especially in severe cases when done early as a primary surgery in congenital glaucoma

Result

To avoid visual loss and amblyopia it is of very much importance to recognize glaucoma in pediatric patients at an early stage. This case fairly draws attention towards the presence of subclinical subtle signs, like in this case enlargement of corneas was the only presentation. Thus it's early recognition and referral by treating pediatricians is of much importance.

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