

Anterior megalophthalmos – A case report

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Abstract: We describe a 10 years old male who suffered from diminution of vision and raised intra ocular pressure (left eye was more severely affected than the right), the use of some ophthalmologic examinations to evaluate the anatomic structures and visual functions of both eyes. Slit lamp and biometric examinations showed very large corneas, very deep anterior chambers, short vitreous length and miotic left pupil. Gonioscopy showed excessive mesenchymal tissue in the angle of the anterior chamber. Corneal transparency, corneal thickness and lens were normal. Because the patient had bilateral megalocornea, raised IOP, deep anterior chamber and atrophic changes in the iris, anterior megalophthalmos was diagnosed. The case was studied in detail as the condition is rare. Secondary effects such as iridodonesis, miosis, atrophy of the iris stroma, lens subluxation and occurrence of cataractous lens should be kept in mind. The importance of biometric data for diagnosis and differential diagnosis in primary infantile glaucoma and other diseases with megalocornea is discussed.

Keywords: gonioscopy, infantile glaucoma, megalocornea, megalophthalmos, raised IOP

I. Introduction

A ten year old male child was brought to the OPD by the mother with chief complaints of blurring of vision, watering from eyes, pain in eyes and colored halos since the last one year. The blurring of vision was gradual in onset and progress. The eyes were noted to be large right from birth by the mother but no treatment was sought. There is no previous history of spectacle use or any other significant history. The father and elder brother of the patient were blind. Both had gradual loss of vision over some months. They were not available for examination.

II. Ophthalmologic Examinations

On Ophthalmologic Examination there was gross obvious enlargement of both the corneas. Thereafter various other Ocular Examinations were undertaken, the findings of the same are as follows:

1. Both the corneas were transparent.
2. Corneal thickness was measured by pachymetry and was found to be near normal.
3. The corneal curvature was measured by keratometry and was found to be normal (K reading: right eye- 43.25/42.75, left eye- 43.00/43.75).
4. The most significant finding in the cornea was the enlarged corneal diameter which was measured with the help of calipers. In the right eye the corneal diameter was 13.5 mm and in the left eye it was 15.5 mm.



Fig. 1



Fig. 2



Fig. 3



Fig. 4

A – Scan Biometry:

Biometry showed the axial length to be increased to 24.93 mm in the right eye and 26.56 mm in the left eye. Also the depth of the anterior chamber was increased in both eyes to 5.68 mm and 6.02 mm for right and left eye respectively. Lens thickness was 3.31 mm for right eye and 3.53 mm for left eye. Vitreous length was 16.6 mm in the right eye and 15.86 mm in the left eye. Vitreous index was 64.82% for right eye and 59.71% for left eye.

Slit Lamp Examination:

Pupil size was 3mm in the right eye and 2mm in the left eye i.e. miotic left pupil. Iris was normal in appearance with the naked eye. Slit lamp examination showed clear corneas, enlargement of the complete anterior segment of the eyes and the left iris showed atrophic patches on the superior and temporal margins. Gonioscopy was done in both the eyes but the angle structures were not clearly visible and could not be well differentiated. There was evidence of excessive mesenchymal tissue in the angle. The intraocular pressure in the right eye was 31.8 mm Hg and in the left eye was 34.4 mm Hg with applanation tonometry. Treatment was started for raised IOP with dorzolamide and timolol maleate 0.5% eye drop. Vision in the right eye was 6/9 and in the left eye was 6/60. Myopic correction was -0.5 D for the right eye and -3 D for the left eye after which vision improved to 6/6 in the right eye and 6/36 in the left eye. Lens and fundus appeared to be normal. On follow up IOP was found to be normal. Patient was continued on anti glaucoma treatment and reviewed every month. Unfortunately chromosomal analysis could not be done for the patient.

III. Discussion

The new born cornea measures about 10 mm in horizontal diameter and reaches the average adult measurement of 11.75 mm by 2 yrs of age. Megalocornea is present if the horizontal diameter of a new born cornea is 12 mm or more and if adult cornea is 13 mm or more.

An enlarged cornea occurs in three patterns:-

1. Megalocornea unassociated with other ocular abnormalities.
2. Anterior megalophthalmos, that consists of megalocornea, iris and angle abnormalities.
3. Buphthalmos in infantile glaucoma¹.

Anterior Megalophthalmos is a rare X linked recessive disease², which manifests as bilaterally symmetrically enlarged clear corneas that remain stable (non progressive) throughout life and sometimes contain a stromal mosaic pattern¹. This condition predisposes the patient to glaucoma³. The patients present a very deep anterior chamber and a vitreous index (vitreous length/axial length into 100) below 69%⁴.

The iridocorneal angle is open but contains excess mesenchymal tissue, whereas iris manifests a hypoplastic anterior stroma and transillumination defects¹. Secondary effects such as iridodonesis, miosis, atrophy of the iris stroma and cataractous lens should be kept in mind².

According to Peter Laibson and George Waring, the two associations that threaten vision are the frequently elevated IOP, which requires annual lifelong examination for early detection, and cataracts which often appear in the fourth decade¹.

If bilateral clear corneas of normal thickness measure 13mm or more in diameter without associated ocular abnormalities, the non progressive disorder of Simple Megalocornea exists and once the diagnosis is clearly made no follow up is necessary¹.

Buphthalmos is a term to describe the abnormal progressive enlargement of an infant's eye secondary to elevated IOP; usually noticed in the first year of life. The classic triad of findings usually seen is epiphora, photophobia and blepharospasm which result from corneal edema, often with associated breaks in the Descemet membrane called Haab striae.

Additional non specific signs of glaucoma in early life include a deep anterior chamber and optic nerve cupping⁵. Also in contrast to megalocornea, congenital glaucoma is marked by the absence of transillumination defects, absence of pigment dispersion and less pronounced enlargement of the anterior chamber.

IV. Conclusion

The differentiation between simple megalocornea, megalophthalmos anterior and primary infantile glaucoma is often difficult, but very important , as in the latter case surgical treatment prevents blindness.

References

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