University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 **2021, Vol. 7(1)**

A case report of Bilateral Megalocornea with Pigment Dispersion Syndrome

Narendran B S
Department of Ophthalmology, Madras Medical College and Government General Hospital

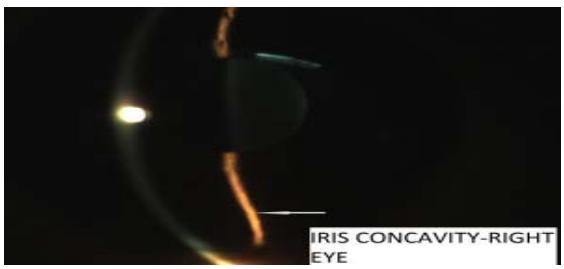
Abstract : A 19 year old male presented with headache and watering in both eyes. Visual acuity in both eyes was 6 by 6. Anterior segment examination of both eyes showed megalocornea, Krukenberg spindle on back of cornea and deep anterior chamber. Measurements showed increase in corneal diameter. Intraocular pressure was normal in both eyes. Gonioscopy showed open angles with trabecular meshwork hyper pigmentation. Ultrasound biomicroscopy revealed iris concavity towards the lens surface suggestive of pigment dispersion syndrome. The patient is on regular follow up to look for signs of development of glaucoma. This case is presented for rare association of megalocornea with pigment dispersion syndrome.

Keyword : Megalocornea, Pigment dispersion syndrome, Krukenberg spindle, Ultrasound biomicroscopy

A 19 year old male presented with headache and watering in both eyes from past 2 weeks. Visual acuity in both eyes 6/6. On ophthalmologic examination there was gross enlargement of both the corneas. Slit lamp examination of both eyes showed normal eyelids, clear conjunctiva, pigment deposition on the corneal endothelium in a vertical spindle-shaped distribution, deep anterior chamber, normal iris colour pattern, 3mm round normally reacting pupils and clear lens. Gonioscopy done in both eyes showed open angles with trabecular meshwork hyper pigmentation. The intraocular pressure in both eyes was 12mm Hg with Goldmann applanation tonometry. Corneal thickness measured by optical pachymetry was found to be 0.405 and 0.419 micrometer in the right eye and left eye respectively. The corrected intraocular pressure for the patient's central corneal thickness was 20mmHg and 19mmHg respectively in the right and left eye. The corneal curvature was measured by keratometry and was found to be normal (K reading: right eye-43.25D/42.75D, left eye-43.00D/43.75D). The most significant finding in cornea was enlarged corneal diameter which was measured with the help of calipers. Corneal diameter was 14.54mm in the right eye and 14.59mm in the left eye. Ultrasound biomicroscopy revealed iris concavity towards the lens surface. Biometry showed the axial length in right eye to be 24.58mm and left eye to be 24.65mm. Anterior chamber was deep measuring 5.07mm and 4.88mm for right eye and left eye respectively. Fundus examination of both eyes revealed a clear media, well defined disc margins, healthy neuroretinal rim with a cup disc ratio of 0.3, vessels with normal thickness and caliber and a positive foveolar reflex.

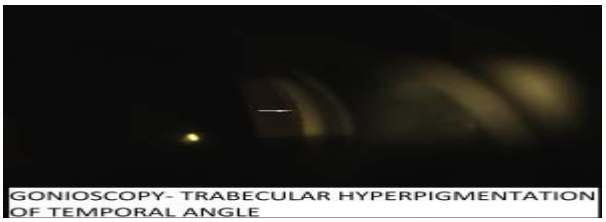
SLIT LAMP EXAMINATION OF RIGHT EYE :

RIGHT EYE- DIFFUSE SLIT LAMP ILLUMINATION

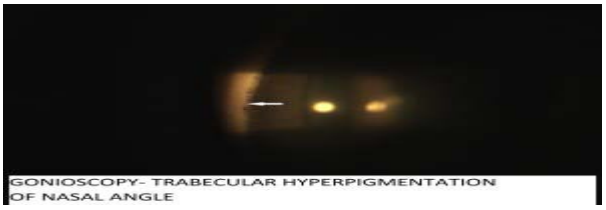


GONIOSCOPY- TRABECULAR HYPERPIGMENTATION OF SUPERIOR ANGLE

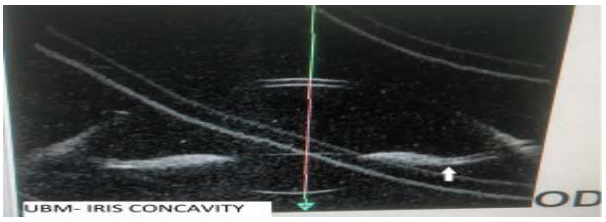
GONIOSCOPY- TRABECULAR HYPERPIGMENTATION OF INFERIOR ANGLE



GONIOSCOPY- TRABECULAR HYPERPIGMENTATION OF TEMPORAL ANGLE



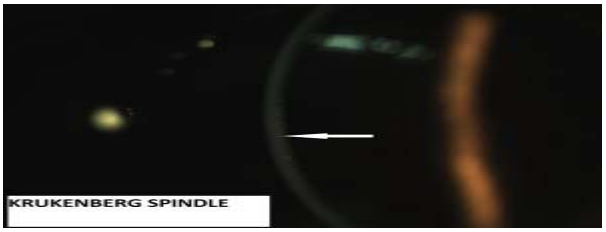
GONIOSCOPY- TRABECULAR HYPERPIGMENTATION OF NASAL ANGLE



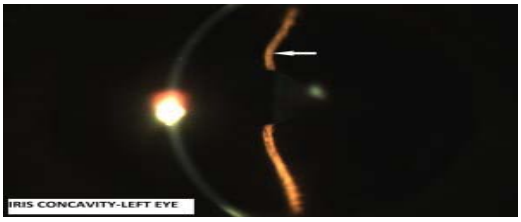
UBM- IRIS CONCAVITY

SLIT LAMP EXAMINATION OF LEFT EYE:

LEFT EYE- DIFFUSE SLIT LAMP ILLUMINATION



KRUKENBERG SPINDLE

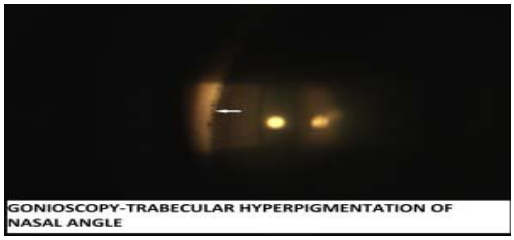


IRIS CONCAVITY-LEFT EYE

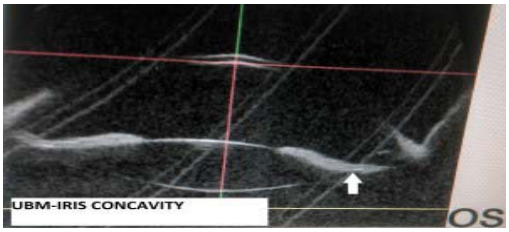
GONIOSCOPY-TRABECULAR HYPERPIGMENTATION OF SUPERIOR ANGLE

GONIOSCOPY-TRABECULAR HYPERPIGMENTATION OF INFERIOR ANGLE

GONIOSCOPY- TRABECULAR HYPERPIGMENTATION OF TEMPORAL ANGLE

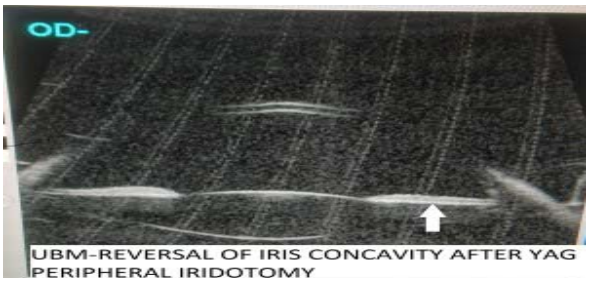


GONIOSCOPY-TRABECULAR HYPERPIGMENTATION OF NASAL ANGLE

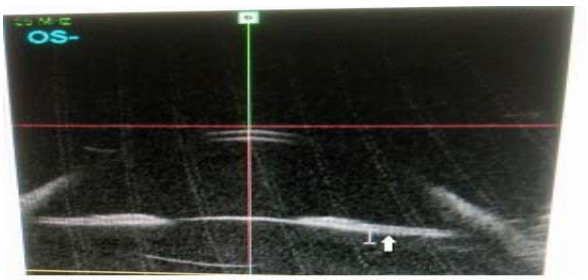


UBM-IRIS CONCAVITY

Patient underwent Nd:YAG LASER peripheral iridotomy prophylactically in both the eyes. After 1 week, patient was reviewed and intraocular pressure was found to be 11mmHg and 12mmHg in the right and left eye respectively by Goldmann applanation tonometry. The corrected intraocular pressure for the patient's central corneal thickness was 19mmHg and 19mmHg respectively in the right and left eye. Anterior segment was quiet, peripheral iridotomy was patent on retro illumination. Repeat ultrasound biomicroscopy was done which revealed reversal of iris concavity



UBM-REVERSAL OF IRIS CONCAVITY AFTER YAG PERIPHERAL IRIDOTOMY



UBM-REVERSAL OF IRIS CONCAVITY

A diagnosis of Both Eye Megalocornea with Pigment Dispersion Syndrome was made based on the above findings. The patient is on regular followup to look for signs of development of pigmentary glaucoma.

Conclusion:

This case is being presented as a rare association of megalocornea with pigment dispersion syndrome.(2,4,5,6) Chin Lin Ho et al has found association of megalocornea with pigment dispersion syndrome.3 This case is being presented to stress importance of screening of patients with megalocornea in their early teens to look for the incidence of pigment dispersion syndrome, such patients if diagnosed promptly can be prevented from developing pigmentary glaucoma by simple intervention like peripheral iridotomy. Studies have demonstrated conversion rates from pigment dispersion syndrome to pigmentary glaucoma to be 11.5 percent at 27 months ,38.8 percent at 4 years and 35 percent at 17 years. (7,8,9) In this case patient presented with both eyes pigment dispersion syndrome and both eyes prophylactic Nd:YAG LASER peripheral iridotomy has been done. After peripheral iridotomy, ultrasound biomicroscopy showed reversal of iris concavity. So patients with megalocornea should be screened and followed up to look for signs of pigment dispersion and once pigment dispersion is seen, it should be managed appropriately and followed up regularly to look for signs of development of pigmentary glaucoma and family members should also be screened.

Discussion

Megalocornea is a rare, bilateral non -progressive condition thought to be due to defective growth of optic cup. Megalocornea is present if horizontal diameter of newborn cornea is 12mm or more and if adult cornea is 13mm or more. Inheritance is X-linked recessive with male preponderance.1 Incidence is 1 in 2000 adults. Physical findings include a dome shaped cornea with normal thickness and occasional central mosaic dystrophy, increased anterior chamber depth, posterior positioning of lens-iris diaphragm, normal intraocular pressure, iris stromal hypoplasia, ectopia lentis, cataract, and pigmentary glaucoma with krukenberg spindles. Patients should be followed regularly for development of glaucoma and cataracts.

References:

1.Miere FM, Dellman JW. Biometry in X linked megalocornea pathognomonic findings: British Journal of Ophthalmology 1994;78:781-85

2.Pigment dispersion syndrome: a clinical study by H G Scheie and J D Cameron British Journal of Ophthalmology 1981 65: 264-269

3.Chin Lin Ho et al. Primary Megalocornea- Clinical features for differentiation from infantile glaucoma. J Pediatric Ophthalmol Strabismus 2004; 41:11-17

4.Cameron W Krukenberg spindle associated with megalocornea and posterior pigmentation of the lens. Am J Ophthalmol 1941; 24687-689.

5.Cavka V. Pigmentary glaucoma. Am J Ophthalmol 1961; 52: 880-894.

6.Sokolic P Megalocornea: report of a case with the signs of pigmentary glaucoma. Am J Ophthalmol 1964; 58486-490.

7.Migliazzo CV, Shaffer RN, Nykin R, Magee S. Long-term analysis of pigmentary dispersion syndrome and pigmentary glaucoma. Ophthalmology 1986;93: 1528-36.

8.Richer CU, Richardson TM, Grant WM. Pigmentary dispersion syndrome and pigmentary glaucoma: a prospective study of the natural history. Arch Ophthalmol 1986;104:211-5.

9.Farrar SM, Shields MB, Miller KN, Stoup CM. Risk factors for development and severity of glaucoma in the pigment dispersion syndrome. Am J Ophthalmol 1989;108:223-9.