



RARE CASE OF ISOLATED ANTERIOR LENTICONUS

Ophthalmology

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KEYWORDS

INTRODUCTION:

Lenticonus is a bulging of the lens capsule and the underlying cortex. The diagnosis of lenticonus is essentially a clinical diagnosis which is made by biomicroscopic examination. According to the localization of the conus a distinction is made between lenticonus anterior and lenticonus posterior. Sometimes isolated, it may be an ocular manifestation of Alport syndrome, a hereditary nephritis accompanied by deafness and other ocular signs such as flecked retinopathy or posterior polymorphous corneal dystrophy. All these manifestations are a result of a genetic defect in type IV collagen, a major component of basal membranes in the human body. The clinical aspects and therapy of anterior lenticonus and Alport syndrome are described, as well as the exploration that must begin when anterior lenticonus is discovered. Preconical lenses are less commonly reported and are more common in Alport syndrome (AS)^[2] If a patient with a conical lens in front of eye is observed clinically, the possibility of being an AS patient should be ruled out first. AS, typically manifested as renal injury (hematuria, proteinuria and progressive renal insufficiency), neurosensorineural deafness and eye injury triad, more common in women, but male patients are more seriously ill, and most of them have renal failure before the age of 40. 11%~92% of AS patients have eye injury, of which anterior conus lens and spotted retinopathy are characteristic changes, mostly involving both eyes, usually only when renal failure occurs. Reference to the Flinter standard or the new Gregory standard. For the family history and eye, ear, and kidney-related examinations of patients with suspected AS, clinical diagnosis can usually be established, and combined with pathological examination and collagen molecular immunopathological examination, clinicopathological diagnosis can be established in most cases.

Case Description :

A 20 year old Asian Indian male presented to our institute with progressive diminution of vision since 8 years in Left eye which has worsened in last 4-5 years. Patient went to various optometrist in past where everytime new glasses were prescribed but unsatisfactory visual improvement achieved. On Ocular examination, (1) refractive error of Right eye was 20/20 and Left eye best corrected visual acuity was 20/120 which was not improving with glasses. (2) Intraocular pressure measured with Goldmann applanation tonometer 15 mmHg of Right eye ; 17 mmHg of Left eye. (3) Slit Lamp examination under dilatation with tropicamide and phenylephrine revealed conical protrusion of anterior surface of crystalline lens suggestive of anterior lenticonus in left eye. (4) There is no abnormality on ocular surface. (5) Axial length measured with USG A scan showed 22.50 mm of right eye 22.13 mm of left eye with AC depth 2.72 mm for the right eye and 2.64 mm for the left eye. (6) Corneal topography showed no obvious abnormalities (6) Keratometry findings of OD 7.84/7.56/11; OS 7.80/7.56/178. (7) Retinal examination with 78 D lens was within normal limit and

Fundus photography: the fundus of both eyes is blurred, part of the retinal blood vessels are faintly out of shape in the shooting area, and the boundaries of the optic disc are clear. Macular OCT shows: the retina of the macular area of both eyes is still flat, and the foveal morphology is visible. The optic disc OCT shows that the thickness of the nerve fiber layer around the optic disc of both eyes was normal. Systemic examination: blood and urinalysis, chest X-ray, electrocardiogram, etc. did not show obvious abnormalities.

Altogether with clinical examination and investigations, diagnosis of isolated anterior lenticonus with lens cataractous changes was made. Patient was informed in detailed about the condition and written, informed consent was taken from the patient for Phacoemulsification with cataract lens aspiration. The surgery went well and patient recovered well. On postoperative day 2, left eye vision was 6/12 with quite anterior chamber. Intraocular pressure was normal. Patient was started on Moxifloxacin and Dexamethasone eyedrops hourly, tobramycin eyedrops hourly, chloramphenicol n polymyxin B eye ointment HS. Patient was discharge next day of surgery and called for followup after one week, 2 week. Left eye vision on Postop day 15 was 6/6.

DISCUSSION-

Lenticonus is a bulging of the lens capsule and the underlying cortex. The diagnosis of lenticonus is essentially a clinical diagnosis was made by biomicroscopic examination. Oil droplet reflex seen on retro illumination. According to the localization of the conus, the distinction between lenticonus anterior and lenticonus posterior.

Lenticonus results from the conical protrusion of the lens anteriorly through the thinnest and weakest part of the capsule. The absence of the a3a4a5 network from the capsule means that it develops partial splits that may rupture. Cataracts develop from healing of small spontaneous ruptures. Lenticonus ceases to progress after cataract formation. Both surfaces of the lens can be affected at the same time but this is very rare. The diameter of the conical protrusion can be between two to seven millimetres.

Lenticonus can be confused with the similar condition, lentiglobus, in which the lens has a spherical protuberance. Patients with lenticonus typically co-present with astigmatism and myopia. In addition to anterior lenticonus, Alport syndrome can manifest as cataracts, corneal arcus juvenilis, posterior polymorphous dystrophy, and flecked retinopathy.

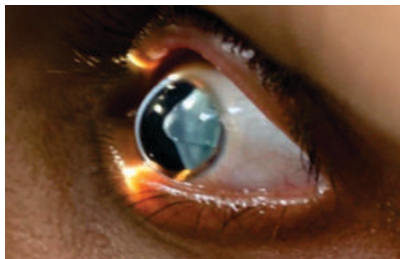
The lens defect may be an isolated finding or more commonly associated with Alport's syndrome. Alport's syndrome is an autosomal recessive disorder with chronic progressive haemorrhagic nephropathy, bilateral high-tone perceptive deafness and ocular findings. Anterior lenticonus, which is virtually pathognomonic of AS, occurs in about 15% of XLAS males and is almost entirely restricted to AS families with progression to end stage renal disease before the age of 30 years and deafness.

Anterior lenticonus is absent at birth, usually appearing during the second to third decade of life, and is bilateral in 75% of patients. Lenticonus worsens until visual symptoms require treatment, and most patients eventually require surgery.

It should be distinguished from amblyopia. Amblyopia is caused by abnormal binocular interaction caused by asymmetric visual input and/or form deprivation caused by congenital or critical period of children's visual development, mostly due to abnormal development of the nervous system, without organic changes in the eye^[1], the vision of adult patients with amblyopia has not been effectively corrected. Treatment for both symptomatic lenticonus and cataract is lens removal and intraocular lens implantation. Lenticonus does not recur after lens replacement.

CONCLUSION –

Anterior Lenticonus is more commonly associated with Alport syndrome. Cataract extraction for these patients is the surgery of choice.



Slit lamp examination: Anterior protrusion of lens capsule.

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