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Original Research Paper

Opthalmology

CASE PRESENTATION: BILATERAL ANTERIOR POSTERIOR LENTICONUS

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ABSTRACT

Aim: To report a case of bilateral anterior and posterior Lenticonus without sensorineural hearing loss, without renal associations. Case report We describe a case of bilateral anterior and posterior Lenticonus and absence of family history. No renal association could be identified on urine routine microscopy and ultrasonography of the abdomen. Clear lens extraction with phacoemulsification with posterior chamber Intraocular lens (PCIOL) was performed for the patient. Conclusion The presence of bilateral anterior and posterior Lenticonus is rare but may be a presentation of Alport syndrome. Clear lens extraction with Intraocular lens is a viable treatment option with good visual prognosis.

KEYWORDS :

INTRODUCTION

Lenticonus is a conical projection of either the anterior or posterior surface of the crystalline lens of the eye, occurring as a rare congenital anomaly. They may be associated with abnormalities of the lens epithelium, by traction from hyaloid remnants, or by localised areas of capsular weakness , which causes bulging. They may be inherited as an autosomal recessive trait or associated with other abnormalities, such as Alport's syndrome or Lowe's oculocerebral syndrome (associated with posterior Lenticonus). They can cause lenticular myopia with irregular astigmatism. Other ocular associations are posterior polymorphous corneal dystrophy, cataract , peripheral thinning, or dot fleck retinopathy.

In the absence of other associated pathologies causing vision loss, clear lens extraction with posterior chamber Intraocular lens is known to give good results . Possible difficulties could be during rhexis due to thin anterior capsule resulting in peripheral run offs on attempting continuous curvilinear capsulorhexis (CCC).

Case report

A 16 year old male presented to the ophthalmology OPD with gradual , painless , diminution of vision in both eyes since 2-3 years . He gives no history of use of glasses in the past. Patient has no history of any eye surgery or history of trauma to the eye. Patient has no history of any decrease in hearing. He has no history of hematuria or proteinuria.

On examination , his unaided vision is 6/36 and 6/18 in right and left eyes respectively. Pinhole improvement was 6/18p and 6/12p of right and left eyes respectively. Patient's autorefractor reading of right eye was -27.25DS, -5.50DC, 20° and of left eye was -27.25 DS, -9.00DC, 170°. Subjective refraction showed no improvement in vision.

On slit lamp biomicroscopy, there was conical protrusion of anterior lens surface in the centre with a distinct 'oil droplet sign' on retroillumination.

On further examination conical protrusion of posterior surface of lens was also seen bilaterally . Both eyes of the patient showed no cataractous changes. Fundus examination of both eyes was within normal limit.

Patient was also referred for pure tone audiometry which was within normal limit. Investigations were ordered to rule out possible renal associations . Serum creatinine was 1.2 mg/dl and urine routine, microscopy was normal. An ultrasonography of the abdomen showed normal findings.

As there was no improvement with refractive correction. So the patient was planned with Right eye phacoemulsification under peribulbar block. Continuous curvilinear capsulorhexis was done with cystitome. Posterior capsule was intact . No complications were faced intraoperatively. Foldable hydrophobic acrylic posterior chamber Intraocular lens was placed in the bag.

Patient had a clear cornea the next day with a vision of 6/9.

DISCUSSION

The lens is surrounded by a lens capsule, which is basement membrane secreted by the capsular epithelial cells and is mainly composed by type IV collagen . Lenticonus is localised bulging of the lens capsule and the underlying cortex that can reach a diameter of 2 to 7 mm; the conus may occur anteriorly or posteriorly . Lenticonus is differentiated by lentiglobus, in which the bulging involves the entire lens surface . On slit lamp, lenticonus is characterised by a transparent, localised, sharply demarcated conical projection of the lens capsule and cortex in axial section . In early stage retro-illumination shows an oil drop configuration. In more advanced stages, associated sub capsular and cortical opacities also appear.

Anterior lenticonus is more commonly seen in Alport syndrome. Alport syndrome, also known as hereditary nephritis is a genetic disorder arising from the mutations in the genes encoding alpha-3, alpha-4, and alpha-5 of type 4 collagen (COL4A3, COL4A4, COL4A5) or collagen 4 □345 network.

The type 4 collagen alpha chains are primarily located in the kidneys, eyes, and cochlea. In 1927, the syndrome of hereditary nephritis and deafness was described by a British physician, A. Cecil Alport. It was observed that hematuria was the most common symptom and males were affected more than females. In 1961, it was named Alport syndrome after having described in multiple family members. It is characterised by renal failure, bilateral sensorineural

hearing loss, and eye abnormalities. Histologic and ultrastructural examination of the kidney in Alport syndrome reveals characteristic, diffuse lamellar ion of the glomerular basement membrane. In the cochlea, there is atrophy of the capillary basement membrane of the stria vascularis . A possible defect in Bruch's membrane has been suggested for the patho genesis of the regional flecks. Eventually, the patients present with proteinuria, hypertension, progressive loss of kidney function (gradual decline in GFR), and end-stage renal disease (ESRD).

Alport syndrome affects about 1 in 50,000 newborns and males are more likely to be symptomatic than females.

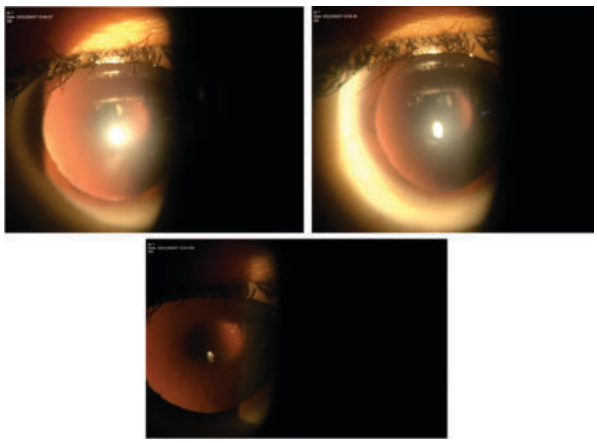
Histopathologic examination shows a thinning of the anterior lens capsule and the presence of vertical dehiscences at the inner part of the central anterior lens capsule. The marked involvement in the anterior polar region may be response to the stresses of accommodation in the area where movement displacement is greatest and the lens is least supported.

Most authors agree that lenticonus posterior is a congenital defect , which is not associated with any systemic disease. Bilateral involvement strongly suggest autosomal dominant inheritance . Amblyopia is the most significant visual problem associated with posterior lenticonus . Amblyopia is caused by optical distortion in the oil droplet stage , by anisometropia or by deprivation due to cataract. Strabismus is also a frequent accompanying defect in children with lenticonus posterior.

The pathogenesis of lenticonus posterior remains unclear. Traction on the posterior lens capsule by remnants of the hyaloid artery system as well as a disturbance in tunica vasculosa lentis has been suggested. In bilateral cases a genetically determined congenital weakness of the posterior lens capsule is likely.

CONCLUSION

The case re-emphasises the simultaneous occurrence of anterior and posterior lenticonus with no systemic associations. Patient has been advised regular follow ups with a nephrologist for possible renal manifestations in future and with the ENT department for any decrease in hearing. Clear lens extraction with IOL implantation improves vision in these patients.



Slit lamp photographs of the patient showing bilateral anterior and posterior lenticonus

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