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A RARE CASE OF PRIMARY IRIS PIGMENT EPITHELIUM CYST

Ophthalomology

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ABSTRACT

An iris cyst is an epithelial-lined space that involves a layer of the iris. Primary iris cysts are nonkeratinized squamous epithelial-lined structures that arise either from the posterior iris layer or stroma. Epithelium from stromal cysts contains goblet cells. The pathogenesis of primary iris cysts is not entirely understood, and proposed mechanisms in the literature are controversial. Iris pigment epithelial cysts are thought to be remnants of the marginal sinus of the iris that have not been obliterated. Primary iris cyst are most commonly of posterior pigment epithelial cell origin and located in the peripheral iris and are often found incidentally. A 69 years old female patient reported to our Outpatient Department (OPD) for regular checkup. Detailed ocular examination with Slit Lamp Bio-microscope revealed a large smooth surfaced, semi-transparent opalescent cystic mass with clear fluid, obscuring the visual axis was seen in the temporal part of the Anterior Chamber (AC) of the left eye. 2x1 mm sized pigmented elevation seen on anterior surface of iris at 6 o'clock angle. The patient was then labelled as a case of Primary Iris Cyst of the left eye. Nd YAG cystostomy was performed under one week peri-procedural coverage of systemic and topical steroids and anti-glaucoma medication. No post-procedural reaction in the AC or rise in IOP was noted.

KEYWORDS

cyst, pigment, iris

INTRODUCTION

An iris cyst is an epithelial-lined space that involves a layer of the iris. A primary iris cyst does not have a recognizable etiology. A secondary iris cyst has a recognizable etiology, such as surgical or nonsurgical trauma.[1]

Classification of Iris Cysts:[2]

- Primary Cysts
- Iris pigment epithelium cysts
- Iris stromal cysts
- Secondary Cysts
- Epithelial downgrowth
- Post-surgical
- Post-traumatic
- Cysts secondary to intraocular tumors
- Iris melanoma
- Medulloepithelioma
- Medication-induced
- Miotics (phospholine iodide)
- Prostaglandins

Primary iris cysts are congenital and occur sporadically. Iris pigment epithelium cysts occur more commonly in adults while iris stromal cysts occur more in children. Primary iris cysts are nonkeratinized squamous epithelial-lined structures that arise either from the posterior iris layer or stroma. Epithelium from stromal cysts contains goblet cells.[3] The pathogenesis of primary iris cysts is not entirely understood, and proposed mechanisms in the literature are controversial. Iris pigment epithelial cysts are thought to be remnants of the marginal sinus of the iris that have not been obliterated. The presence of goblet cells in iris stromal cysts suggest that they are formed by entrapped surface ectoderm tissue likely during formation of the lens vesicle (4th week of embryogenesis).[4][5][6]

Primary iris cyst are most commonly of posterior pigment epithelial cell origin and located in the peripheral iris.[7] Iris cysts are often found incidentally. Though they may be congenital, they may not be detected until late into the first or second decade of life. Patients may notice a "spot" on their iris.[8] Rare reports exist in the literature describing an inheritance pattern of iris cysts, which may present as multiple or bilateral iris cysts in successive generations.[9]

Depending on the size, location and compression of adjacent structures, the following complications may occur secondary to iris cysts:

- Angle-closure glaucoma
- Plateau iris syndrome
- Pigment dispersion syndrome
- Corneal edema
- Focal cataract
- Lens subluxation
- Iritis

CASE STUDY

A 69 years old female patient reported to our Outpatient Department (OPD) for regular checkup. There was no history of redness, flashes, floaters or diplopia. She did not give any history of ocular injury, topical medication or any history suggestive of neurological illness. There was no history of previous ocular surgery.

Ocular examination revealed a visual acuity of 6/18 in right eye and 6/6 in left eye. There was no bony deformity in the orbital margins. Lids and adnexa were also normal. She was orthophoric with ocular movements full and free in all cardinal directions of gaze. Detailed ocular examination with Slit Lamp Bio-microscope revealed a normal right eye. A large smooth surfaced, semi-transparent opalescent cystic mass with clear fluid, obscuring the visual axis was seen in the temporal part of the Anterior Chamber (AC) of the left eye. 2x1 mm sized pigmented elevation seen on anterior surface of iris at 6 o'clock angle. There was no change with decubitus suggesting that the cyst was fixed. The pupil was pushed posteriorly by large iris cyst. There were no features suggestive of previous ocular injury or surgery. Fundus examination could not be done.

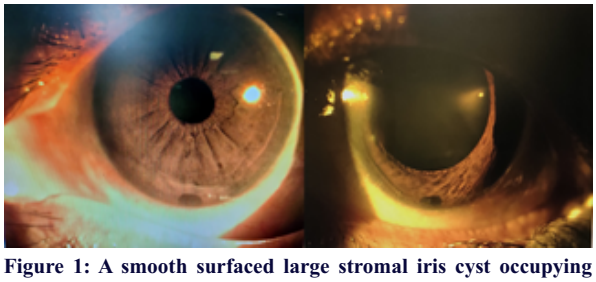

Figure 1: A smooth surfaced large stromal iris cyst occupying anterior chamber.

Ocular ultrasound AB scan did not reveal any abnormality in posterior segment in both the eyes. Ultra Bio-Microscopic scan (UBM) done revealed a smooth, thick walled cystic swelling arising from the iris and occupying the temporal part of the anterior chamber. The lumen of the cyst was anechoic. MR scan of the brain and orbit to rule out any similar lesion was non-contributory. There was no rise in intraocular tension. Routine blood examination and blood sugar level were normal. Microscopic examination of the stool on three consecutive days was also normal. The patient was then labelled as a case of Primary Iris Cyst of the left eye. Nd YAG cystostomy was performed under one week peri-procedural coverage of systemic and topical

steroids and anti-glaucoma medication. No post-procedural reaction in the AC or rise in IOP was noted for one week. Last follow up of the case has shown no recurrence.

DISCUSSION

Iris cysts have been classified into primary and secondary types. Primary iris cysts which commonly arise from the posterior pigment epithelium of the iris are more common in children as compared to secondary iris cysts. In a large series of 251 cases of iris cysts in children less than 20 years of age, primary and secondary iris cyst have been reported in 53 and 4 cases respectively. Of the 53 primary iris cysts, 44 were pigment epithelial and nine were stromal. The peripheral pigment epithelial cyst accounted for 34 cases or 59% of all childhood iris cysts. They are more common in girls and most commonly diagnosed in the teenage years. The primary iris cysts are benign, usually stationary or even regress in some cases and do not require any intervention.[10] However very rarely they enlarge, occupy anterior chamber and have a thick-walled clear lumen. When they obscure pupillary axis or cause pressure symptoms, surgery or by Laser cystostomy as done in our case is required. [10-12] An adequate steroid cover and anti-glaucoma measure should be considered to prevent any Anterior Chamber reaction.

Secondary iris cysts develop due to trauma, malignancies such as melanoma or medulloepithelioma and long-term topical instillation of pilocarpine and/or prostaglandin analogues in the management of glaucoma. Parasitic infestation such as echinococcosis and cysticercosis may also sometimes present as cysts in anterior chamber of eye. In addition to complete removal of iris cyst, iridocyclectomy, lensectomy, vitrectomy, penetrating keratoplasty and intraocular (IOL) removal has been documented and may be needed.[13] Care is needed to prevent any spillage of the contents of the cysts to avoid toxic reactions.[14-15]

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