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Case report

Cogan-Reese syndrome with Iris cyst: A novel presentation

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ABSTRACT

Purpose: To report an atypical case of Cogan-Reese syndrome associated with iris cyst in a young adult male.
Methods: Slit-lamp biomicroscopic examination, swept-source anterior segment optical coherence tomography (ASOCT) and ultrasound bio-microscopy (UBM) were done to evaluate and characterize the nature of the iris cyst. Gonioscopy, specular microscopy and confocal microscopy were attempted, but unsuccessful due to the large corneal opacity.
Results: On slit-lamp biomicroscopy, a large nasal corneal opacity with overlying band-shaped keratopathy was noted, with history suggestive of a trivial non-penetrating trauma and likely healed corneal ulcer. Through the temporal clear cornea, the iris displayed altered pattern with overlying shiny membrane and multiple, small, discrete, hyperpigmented, irregular nodules suggestive of Cogan-Reese syndrome. On the nasal side, an iris cyst with typical 'stuck-on appearance' onto the endothelium was visible. ASOCT and UBM failed to show any evidence of epithelial downgrowth or Descemet membrane disintegrity, ruling out the possibility of a post-traumatic implantation iris cyst.
Conclusion: The occurrence of iris cyst in this case of Cogan-Reese syndrome is unique, and could be related to the disease pathogenesis, or a rare co-incidental finding.

1. Introduction

Iridocorneal endothelial (ICE) syndrome is a spectrum of disease characterized by unilateral, progressive anterior segment morphological changes due to altered 'epitheloid-like' endothelial cells, called the ICE cells [1]. This change in endothelial cell characteristics results in corneal decompensation, and its progress over the angle and iris surface in the form of a proliferative contractile membrane results in intractable glaucoma and iris configurational changes [2]. Depending on the chief manifestation, the spectrum varies from Chandler syndrome, mainly affecting the cornea, to Cogan Reese syndrome and Essential iris atrophy where the iris features predominate. It is sporadic in nature, with the aetiology being speculated to be a precedent viral infection [2]. They have a female predilection, and are known to occur in young to middle aged adults.

Cogan-Reese syndrome (CRS), also called the 'Iris Nevus syndrome', has characteristic iris nodules arising due to pinching of the iris stroma by the overlying aberrant translucent endothelial membrane. Common associated features include loss of normal iris pattern, corectopia, iris heterochromia and corneal edema [2]. This report presents a case of CRS associated with band shaped keratopathy (BSK) and iris cyst in a young male; such atypical presentation being the first of its kind.

2. Case summary

A 19-year-old male presented to us with complaints of left eye (LE) white opacity for the last 9 years, which slowly increased in size, with associated painless diminution of vision since then. He gave history of a preceding trivial non-penetrating trauma with gravel, followed by redness, pain and discharge which got better with topical medications. A white opacity had developed thereon.

On examination, the best corrected visual acuity (BCVA) RE was 6/6 and LE was finger counting close to face with accurate projection of rays. There was a left divergent squint of approximately 30° on Hirschberg test. Slit-lamp biomicroscopic examination revealed an inferonasal 6*7 mm irregular white corneal opacity with overlying band-shaped keratopathy. A slit beam through the peripheral part of the opacity suggested associated superficial stromal scarring, while the central area being dense, precluded depth assessment. The temporal part of the cornea seemed to be normal.

Just above the opacity, a brown mass was seen posterior to the cornea, with typical 'stuck-on appearance' onto the endothelium

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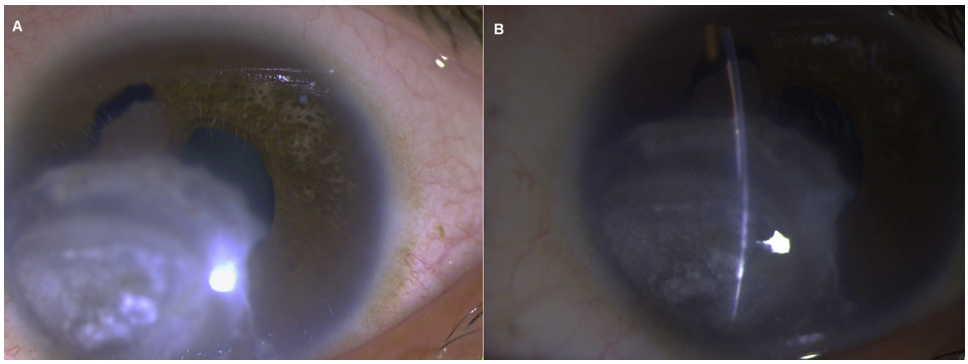

Fig. 1. (a) Slit-lamp diffuse illumination photograph showing inferonasal corneal opacity with band shaped keratopathy. Note the 'stuck-on appearance' of the iris cyst to the endothelium above the opacity. Shadow of the mass appreciable posteriorly due to its elevation. On supero-temporal iris, multiple hyperpigmented discrete typical Cogan-Reese nodules are appreciable with corectopia. (b) Slit-lamp direct focal illumination showing band shaped keratopathy over the corneal opacity and superior iris cyst stuck onto the endothelium.

(Fig. 1). Indirect focal illumination showed the mass to be cystic in nature, with clear contents. Being pigmented and appearing to arise from the iris surface and secondarily impinging on the endothelium, a diagnosis of iris cyst was made. However, the cause for the cyst merited investigation. The temporal iris visible through the clear cornea showed incidental altered iris pattern with overlying shiny membrane and multiple, small, discrete, hyperpigmented, irregular nodules over a focal area along with corectopia (Fig. 1). The other parts showed lack of normal iris pattern and pupillary ruff atrophy. A dilated fundus examination revealed a healthy disc and macula. IOP on tonopen was 16 mmHg. The large corneal opacity precluded gonioscopy. RE was within normal limits.

Multimodal imaging with swept-source anterior segment optical coherence tomography (ASOCT) (SS-1000, CASIA, Tomey Corporation, Nagoya, Japan) and ultrasound biomicroscopy (UBM) (Vumax, Sonomed Escalon, New York, USA) was undertaken to assess the depth of corneal opacity and define the iris cyst (Figs. 2 and 3). Both of them showed corneal opacity to involve only the superficial stroma. There was no evidence of any epithelial downgrowth, and the Descemet membrane was intact in all the sections ruling out the possibility of a penetrating corneal trauma, and hence eliminated the likelihood of a post-traumatic implantation iris cyst. On ASOCT, the cyst wall was seen to be in continuum with the iris surface, and a clear line of demarcation was seen between the anterior cyst wall and thickened Descemet membrane (Fig. 2, white arrow). Specular imaging and Confocal microscopy were attempted in the temporal clear cornea but were unsuccessful.

Thus, a clinical diagnosis of Cogan-Reese syndrome with iris cyst was made, which showed associated corneal opacity and band shaped keratopathy. The patient having a divergent squint and stimulus deprivation amblyopia, the poor visual prognosis was explained and advised regular follow-up due to the risk of future glaucoma.

3. Discussion

The term "Iridocorneal endothelial" syndrome was given by Yanoff in 1979 [3]. It unified the features of essential iris atrophy described by Harms, the eponymous Chandler syndrome and the iris nodules described by Cogan and Reese.

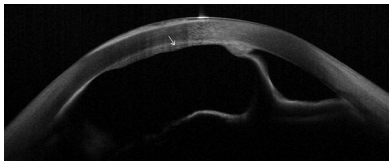

Fig. 2. Swept source anterior segment optical coherence tomography showing corneal opacity to involve up to the superficial stroma only. The cyst is in continuum with the iris posteriorly, and anteriorly impinges over the endothelium causing overlying Descemet thickening and scarring. A clear line of demarcation is seen between the cyst wall and the endothelium (white arrow).

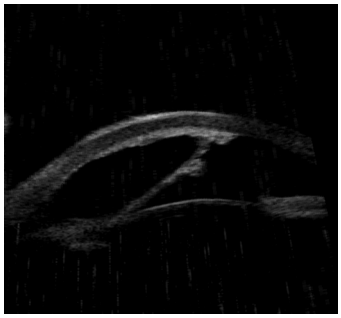

Fig. 3. Ultrasound biomicroscopic image showing the corneal opacity to involve up to the superficial stroma only. The mid and posterior stroma appear normal. The cyst is in continuum with the iris posteriorly, and anteriorly impinges over the endothelium causing overlying Descemet thickening and scarring.

Cogan-Reese syndrome predominantly affects the iris and may present with myriad manifestations such as iris nevus, peripheral anterior synechiae, matted appearance of iris, loss of iris crypts, fine iris nodules, ectropion uveae, heterochromia and secondary glaucoma [1,2,4]. Iris cyst in a case of Cogan-Reese syndrome is being reported for the first time in literature. A single case of band-shaped keratopathy (BSK) due to dysfunctional endothelium of the ICE syndrome has been reported previously [5]. The present case has a large dense BSK, and with antecedent history of trauma and probable healed corneal ulcer being the mostly likely cause [6], there could be a possibility that the ICE syndrome hastened/increased its severity. Also, it could have got aggravated due to the circumscribed contact of the iris cyst onto the endothelium, causing localised endothelial compromise with Descemet membrane thickening and scarring.

The differential diagnosis for multiple pigmented iris nodules include Lisch nodules of neurofibromatosis, tapioca melanoma, diffuse malignant melanoma, iris mammillations of oculodermal melanocytosis, inflammatory nodules associated with granulomatous uveitis, metastatic seedlings and ICE syndrome [2,7]. Among these, this patient best fitted into the diagnosis of ICE syndrome (Cogan-Reese variant), it being unilateral with classic shiny translucent membrane and focal hyperpigmented nodules in the absence of any active ocular inflammation or systemic disorder. However, the presentation of iris cyst in a case of ICE syndrome is novel, having ruled out the possibility of post-traumatic implantation cyst [8]. It could be a true iris cyst, or a pseudo-cyst due to lifting up of the proliferated endothelial membrane from over the iris surface carrying its pigments, resulting in a pseudocyst wall. With respect to the incidence of glaucoma, a study comparing the features of all three variants of ICE syndrome showed that up to 37% of CRS patients had a cup-disc ratio of < 0.5. However, when affected, the severity was found to be worse [9].

In conclusion, ICE syndrome is a progressive disease with unclear pathogenesis and heterogenous manifestations. This report features the

occurrence of iris cyst in a case of CRS, that could be related to the latter's pathogenesis, or a mere co-incidental finding.

Authors statements

All the authors have read and approved the final manuscript for submission.

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Conflict of interest

None declared for all the authors.

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