



A RARE CASE OF KERATOMALACIA LEADING TO SPHEROIDAL DEGENERATION AND GLAUCOMA

Ophthalmology

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ABSTRACT

Spheroidal degeneration is acquired, non-hereditary corneal degeneration predominantly affecting males over forty. Characterized by bilateral, usually asymmetrical opalescence of anterior layers of cornea, two forms are reported viz. primary and secondary. Whereas primary form is an ageing process affecting only peripheral cornea, the secondary form follows other ocular diseases and is probably multifactorial associated with microtrauma. We hereby report a rare case of bilateral secondary spheroidal degeneration of cornea following keratomalacia, precipitated by measles, at the age of two years and associated with bilateral glaucoma who presented at 26 years of age. Cause of glaucoma still remains a mystery.

KEYWORDS

spheroidal degeneration, keratomalacia, measles, glaucoma

INTRODUCTION

Spheroidal degeneration is characterised by bilateral, globular yellowish oily deposits limited to anterior cornea. These may be associated with subepithelial vascular invasion and perilimbal conjunctival deposits. These globules stain positively with haematoxylin, congo red but do not show birefringence with polarised light and are autofluorescent under UV illumination of unstained sections.^[1,2] Pathogenesis is probably multifactorial associated with microtrauma.^[1,2,3] Two basic forms have been reported viz. primary and secondary. Primary form is an ageing process affecting only peripheral cornea and generally without visual implications. Secondary form is associated with other ocular pathology like absolute glaucoma, phthisis bulbi, post-traumatic scars, lattice corneal dystrophy.^[4] Uretts Zavalia have described three stages of the disease.^[1]

CASE REPORT

A 26 year woman presented with episodic unilateral left sided headache with redness and nausea since the age of 11 years. She had contracted measles at the age of two followed by development of whitish deposits in both the eyes Left > Right associated with progressive diminution of vision. Born at full term by normal vaginal delivery, did not require resuscitation and attained normal milestones but had no immunization or vitamin A prophylaxis. At presentation, best corrected visual acuity (BCVA) in right eye (R/E) was 6/12 and PL denied in left eye (L/E). Both corneal diameter were 11.5X10.5mm in horizontal and vertical dimension. Right eye had crescentic fleshy mulberry gelatinous growth from 2.30 to 6.30 o'clock involving peripheral 1/3rd of cornea with overlying superficial vascularization and adherent leucoma at 2.30, 4 and 6:30 o'clock (Figure 1a). Left eye had similar central 8X10mm growth with superficial vascularization at 7, 11 and 12 o'clock. Additional soapy deposits were present as clumps at 10 and 5-6 o'clock (Figure 1b). Corneal sensation was reduced in involved areas. Intraocular pressure (IOP) was 46mm Hg in R/E and digitally very high in L/E. Gonioscopy in right eye revealed high iris insertion with no identifiable angle structures in any quadrant with iridocorneal adherence limited to the region of lesion (Figure 1c) i.e. approximately 120°. Axial length was 19.81mm and 26.74mm in right and left eye respectively (from surface of growth).

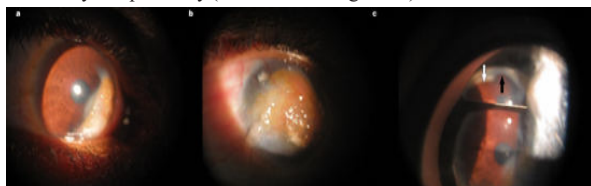


Figure 1a,b: Slit lamp photograph of right and left eye showing fleshy mulberry gelatinous growth with overlying yellowish crystalline deposits and superficial vascularization. Notice the soapy deposits at 10 and 5-6 o'clock in left eye.

Figure 1c: Gonio-photo of inferior angle of right eye showing high iris insertion with no identifiable angle structures (white arrow) and iridocorneal adherence in the region of lesion on nasal side (black arrow).

Fundus evaluation of right eye showed cup:disc ratio of 0.6:1 with baring of both superior and inferior circumferential vessels (Figure 2a). Ultrasound of left eye showed excavation cupping of disc (Figure 2b).

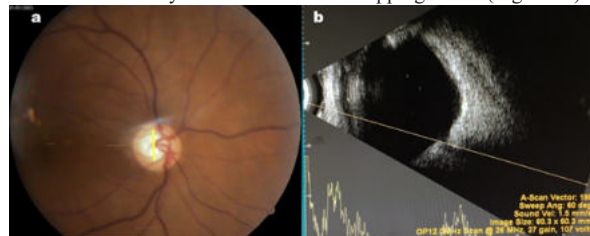


Figure 2a: Color fundus photograph of right eye showing a cup:disc ratio of 0.6:1 (yellow streak) with baring of both superior and inferior circumferential vessels

Figure 2b: Ultrasound scan of left eye showing excavation cupping of optic disc

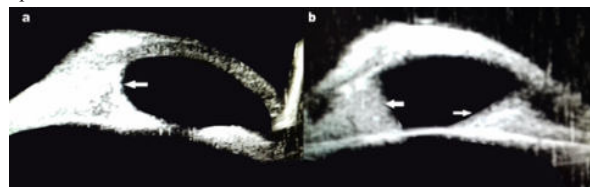


Figure 3a, b: Ultrasound biomicroscopy (UBM) of a) right eye, b) left eye showing the lesion limited to the cornea as well as areas of iridocorneal adherence (white arrows)

Ultrasound biomicroscopy (UBM) showed the lesion to be limited to cornea in both eyes with areas of iridocorneal adherence (Figure 3a,b). Incision biopsy of lesion from left eye showed hyperplastic stratified squamous epithelium lined tissue with numerous spheroid shaped hyaline deposits scattered in epithelium and sub-epithelium (Figure 4a,b). Deposits were autofluorescent (Figure 4c), positive for Congo red and did not polarize.

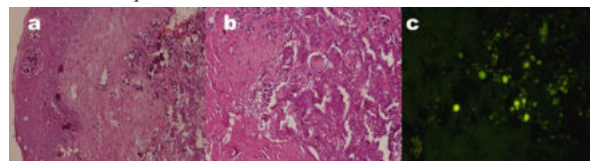


Figure 4a,b,c: Histopathology photographs (10X) of left eye; showing hyperplastic stratified squamous epithelium lined tissue (a) with numerous spheroid shaped hyaline deposits seen scattered in sub-epithelium and overlying epithelium (b) Autofluorescent deposits as seen under UV illumination of unstained sections (c)

They were negative for Colloidal Iron, Von Kossa, Masson Trichrome, Methyl Violet and PAS stains. Features were suggestive of spheroidal degeneration. Patient was managed with anti-glaucoma drugs in right eye and lubricating eye drops in both eyes and prognosis was

explained. Cyclo-cryotherapy in left eye was done for painful blind eye. Target pressure of 15mm Hg was achieved in right eye with topical Latanoprost 0.005% and Timolol 0.5% with follow up of 2years. The pain in left eye reduced considerably post cyclo-cryotherapy.

DISCUSSION

This patient is a case of secondary spheroidal degeneration as proven histopathologically. She is from low socio-economic strata, never took milk and has neither been immunised for any disease nor has taken vitamin A prophylaxis. This made us presume that she had vitamin A deficiency in her childhood and measles precipitated development of keratomalacia. Keratomalacia led to bilateral corneal perforations which resulted in formation of adherant leucomas and secondary spheroidal degeneration. Presence of glaucoma has been of reasonably long duration as evidenced by excavation of disc in left eye and very high IOP in right eye. However, it must have happened after the age of 3-4 years as corneal diameters are normal. Furthermore, moderate cupping with very high pressures of 46 mm Hg is not compatible with the presence of glaucoma of a very long duration. The presence of anterior iris insertion on gonioscopy, glaucoma development years after corneal perforation and very high IOP with 2/3rd of angle functioning normally go in favour of primary juvenile (developmental) glaucoma rather than that secondary to the disease process, although the same cannot be ruled out. This case gives us food for thought to look for any relationship between spheroidal degeneration and juvenile in etiopathogenesis, progression or prognosis of disease. Cause of poor visual acuity in right eye appears to be high astigmatism due the lesion and that in left eye is due to involvement of pupillary axis by lesion as well as advanced glaucoma. Presence of iridocorneal adhesion is limited to certain areas with normal AC depth in between as seen on UBM, which suggests corneal perforation as a cause of the adherence rather than the disease itself. As far as management options are concerned, if the IOP is not controlled medically in right eye, filtering surgery can be undertaken like any other case. However, cyclodestructive procedures were a viable option in left eye in view of painful blind eye. Keratoplasty in either eye is not indicated for optical/cosmetic reasons as in right eye the glaucoma might worsen with keratoplasty and in left eye, due to advanced glaucoma and likelihood of recurrence of pathology in the graft as well. The features which make this case unique include: there is neither a reported association of spheroidal degeneration with keratomalacia or with measles nor with juvenile glaucoma.

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