Case Report

Cholesterosis Bulbi in a Painful Blind Eye with High Intraocular Pressure and Long Standing Total Retinal Detachment

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This is case report of a 19 year old male who presented to the Ophthalmology Department of Jinnah Hospital Lahore in July 2011 with a painful blind left eye for the last two years. Examination of left eye showed no perception of light, circumcorneal injection, band keratopathy, pseudohypopyon of polychromatic crystals, polychromatic crystals embedded in iris stroma and aphakia. Intraocular pressure was 32 millimeters of Mercury. B scan ultrasound showed old retinal detachment. Right eye examination was normal. There were no associated systemic examination findings. He had a history of cataract surgery for left congenital cataract at the age of four years followed by sudden painless loss of vision three years after cataract surgery. In our eye department, he was started on topical cycloplegics, corticosteroids and topical and systemic anti-glaucoma medication and was made symptomatically comfortable. Retinal surgery was not contemplated taking into consideration his chronic retinal detachment and poor visual status.

Cholesterosisbulbi is a condition involving presence of polychromatic, white or golden crystal in the vitreous cavity and / or anterior chamber. This condition is also known as hemophthalmos or synchysis scintillans[1]. This condition typically occurs as a sequel of chronic vitreous hemorrhage[2] but may occur in cases of long standing retinal detachment, ocular trauma and advanced Coats disease[3,4]. Cholesterol crystals in anterior chamber is a rare manifestation of this condition, which may be found in advanced cases of cholesterosis bulbi[3,5].

These crystals are composed of cholesterol which is derived from degradation products of red blood cells or plasma cells. They can be found freely or engulfed within foreign body giant cells[2]. In addition these crystals can also form from breakdown of vitreous and from subretinal fluid of a long standing retinal detachment[6]. In anterior chamber, these crystals can be found in anterior chamber angle, embedded on iris or may form a hypopyon. In vitreous cavity, these crystals are found suspended in vitreous which tend to settle inferiorly when the eye is immobile.

A considerable number of cases with Cholesterosisbulbi have been treated with enucleation due to intractable pain associated with it and the risk of sympathetic ophthalmitis in the other eye.[7] We are here to report the first case of Cholesterosisbulbi in Jinnah Hospital Lahore, associated with profound involvement of anterior chamber with cholesterol crystals, concurrent increased intraocular pressure and long standing total retinal detachment.

CASE HISTORY

A 19 year old male was brought to Outdoor Patient Department of Ophthalmology Unit 1 in Jinnah Hospital Lahore in July 2011 with the primary complaints of painful and blind left eye for last 2 years. His past ocular history revealed surgery on his left eye for congenital cataract at the age of 4 years (15 years ago) from an eye clinic at Chakwal, Punjab. Patient was left aphakic after primary surgery. He was prescribed aphakic spectacles for visual correction. After 3 uneventful years, the patient suffered from sudden painless and severe decline in visual acuity which converted to no perception of light after few

months of no intervention. He had no history of trauma and was not using any ocular or systemic medication at the time of his presentation to us. Family history was also insignificant. The patient did not have any medical record available for his previous ocular treatment.

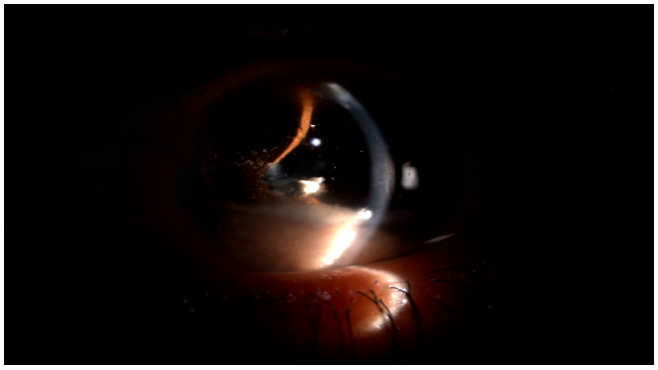

Fig 1: Anterior chamber photograph showing pseudohypopyon of polychromatic crystals and shallow anterior chamber.

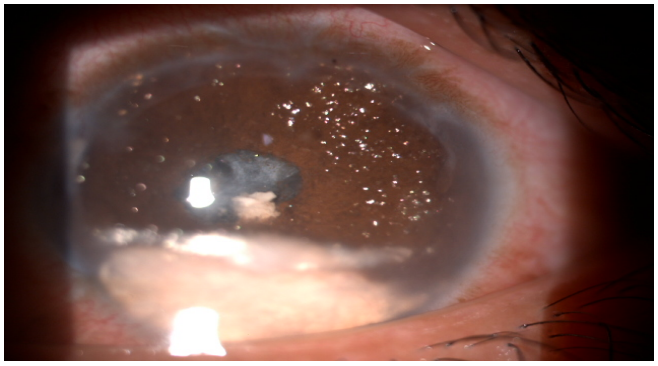

Fig 2: Anterior chamber photograph showing cholesterol crystals embedded on iris surface.

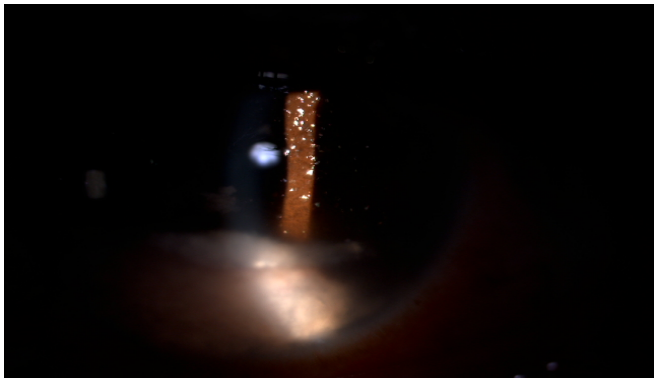

Fig 3: Anterior chamber photograph showing cholesterol crystals embedded on iris surface.

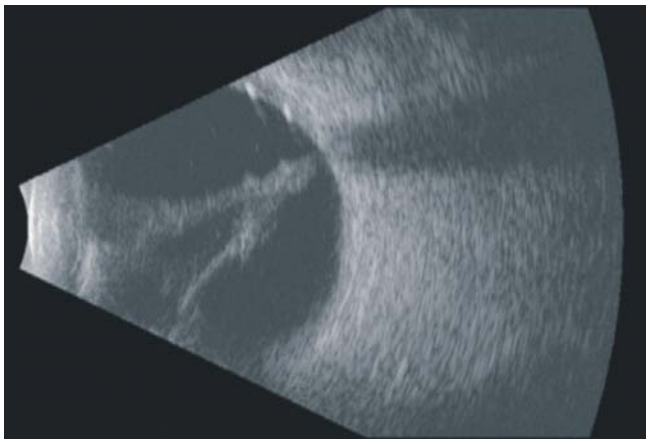

Fig 4: B-Scan showing total retinal detachment.

Examination of the eyes showed normal right eye and no perception of light in his left eye. Anterior segment examination of the left eye showed circum-corneal injection, band keratopathy, shallow anterior chamber, pseudohypopyon of polychromatic crystals measuring 3 – 4 mm (Fig 1), polychromatic crystals deposited on iris stroma (Fig 2, 3), grade 1 flare and +2 cellular anterior chamber activity, interrupted posterior synechie with non reactive 3mm roughly round pupil and strongly positive reverse Marcus Gunn reaction, aphakia with intact but thickened and opacified posterior capsule. Gonioscopy revealed cholesterol crystals in anterior chamber angle. Goldmann's tonometry displayed intraocular pressure of 14 and 32 millimeters of mercury in right and left eye respectively. There was no view available for examination of vitreous and retina. B scan ultrasonography showed left sided total retinal detachment (Fig 4).

His rest of general and systemic examination was unremarkable.

No retinal surgery was advised to the patient, he was started on topical Atropine 1% three times a day, topical Dexamethasone 0.1% four times a days, topical Timolol maleate 0.5% two times a day and oral Acetazolamide 250 mg four times a day. He was also advised protective polycarbonate glasses for his right eye and to avoid contact sports. He was asked to follow up after 3 days of initial visit. At his first follow up, he was found to have intraocular pressure of 27 millimeters of Mercury with mild reduction in his ocular symptoms. After a month of regular treatment and follow up and with addition of topical Brimonidine tartrate 0.2% three times a day, his intraocular pressure was successfully controlled to 18 millimeters of Mercury with occasional cells in

anterior chamber. The patient was also noted to have significant improvement in his ocular symptom. Currently he is on 15 day follow up with our department.

DISCUSSION

Cholesterol crystals have been demonstrated in most tissues of eye but the commonest sites include lens, vitreous and retina. They usually occur as a long term consequence of ocular trauma, inflammation of uveal tract, degeneration, particularly of vitreous; and rarely neoplasia[7]. In a number of cases, the eye has been blind for a number of years and these crystals have been found accidently in anterior chamber. Suresh[7] conducted microscopic examination on these crystals and found them to be composed of cholesterol in the form of thin colorless transparent plates of square or rectangular shape. Stevens calculated the normal concentration of cholesterol in normal aqueous and found it to be considerably lower than plasma cholesterol levels. He also demonstrated the chemical nature of these crystals through chromatography to be cholesterol[8].

The major source of these cholesterol crystals has been identified to be degenerating red blood cells either from hyphaema or vitreous hemorrhage[9]. Long standing intraocular inflammation resulting in defective blood retinal barrier can also result in extravasation of cholesterol in the eye and thereafter, its deposition in different ocular tissues; aphakia is also a recognized cause of deposition of cholesterol crystals in anterior chamber[6]. Kennedy[6] also reported cases of cholesterosisbulbi involving anterior chamber resulting after long standing retinal detachments with no evidence of intraocular hemorrhage as reported in our case. Forsius[4] believed that the process the deposition of cholesterol accelerates when there is clinically demonstrable evidence of intraocular inflammation because proteins and fats enter the chamber with the flow of fluid in the eye, and as we know that cholesterol is insoluble in water, it crystallizes. An important factor in the deposition of crystals seems to the time for which the eye has remained blind[7]. All the seven cases reported by Forsius[4] had been blind for more than 5 years and in Gruber's series, atleast 6 cases had no sight for more than 5 years. Awan[10] reported a case of cholesterol crystals in anterior chamber of a 15 year old white girl with a structurally and functionally normal eye.

The cause for high intraocular pressure can be secondary to deposition of cholesterol crystals in anterior chamber angle or direct damage of trabecular meshwork by the crystals[3]. This was the suspected reason for raised intraocular pressure in our case, since gonioscopy did not reveal any other angle pathology apart from cholesterol crystals in angle. Under these circumstances the eye can be made comfortable by conservative measures as shown by Kumar[7]. This was the mainstay of treatment in our patient. In the case reported by Park[3], the causative factor for high intraocular pressure was neovascularization in anterior chamber angle, which was successfully treated by intravitreal injection of Bevacizumab along with pars planavitrectomy.

In the past, the mainstay of treatment in patients with painful blind eyes along with cholesterosisbulbi has been enucleation, mainly due to ineffective treatment available and potential risk of sympathetic ophthalmitis[9,11]. With advances in therapeutic ophthalmology in the form of better anti-glaucoma therapy, potentially more effective anti inflammatory medications, lasers and anti-vascular endothelial growth factor agents, a more conservative and cosmetically acceptable approach has been adopted for such cases. Such cases need to be in a close follow up so that additional and alternative treatment can be offered in the event of recurrent or persistently uncomfortable eye.

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