



Salzmann's Nodular Degeneration Right Eye – A Case Report

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Introduction

Salzmann's Nodular Degeneration is a condition where a "callus" like nodule forms on the front surface of the cornea. It is a slowly progressive condition in which gray-white to bluish nodules measuring 1-3 mm are seen anterior to Bowman's layer of the cornea, usually bilaterally^{1,2,3}. These elevated nodules can be located near the limbus or in the mid-peripheral cornea⁴. We report a case of 50 year old male diagnosed with Salzmann's degeneration right eye.

Case Report

A 50 years old male reported in ophthalmology OPD of a tertiary care center of North India with a 3-year history of painless, progressive loss of vision in both eyes. The diminution of vision was more in right eye in comparison to the other eye. It was associated with photophobia, itching and foreign body sensation in right eye. The patient was seeking treatment of blepharitis diagnosed elsewhere. He had three updates of his spectacle prescription that had not provided satisfactory vision. There was no diurnal variation in vision. There was no other history of trauma, exposure of

toxic materials to the eye, use of contact lenses or any systemic illness.

The visual acuity was 5/60 unaided in right eye and 6/24 left eye. On slit lamp examination there were multiple flakes present on the eyelashes of both eyes and right eye revealed waxy secretions over the lid margins. Greyish white nodules were present on the cornea between the corneal epithelium and Bowman's layer. They were located at the paracentral region extending centrally involving the pupillary area (Fig 1). Rest of the ocular examination including the fundus was within normal limits. The Schirmer's test was done to rule out dry eye. The values read 28 mm in right eye and 32mm in left eye. All the signs suggestive of exposure keratopathy were ruled out like any lid abnormality, proptosis, lagophthalmos, decreased blink frequency and weakened orbicularis strength which are typical features of Bell's palsy.

The patient was diagnosed with Salzmann's nodular degeneration based on characteristic presentation of nodules on slit lamp examination. There was progressive deterioration of vision and progressive increasing irregular astigmatism.

The patient was planned for superficial keratectomy. He was started on oral and topical antibiotics to reduce pre operative infection and treatment was prescribed for meibomitis and blepharitis. Immediately prior to removal of nodules under the microscope, slit-lamp examination was done to identify the location and extent of the nodules.

The patient was prepped and topical anesthetic was administered in right eye. Epithelial debridement was performed manually using a blade and with assistance of alcohol. Forceps were used to grasp the edge of the nodule firmly to raise the edge. Once the plane of dissection was visible, the nodule was peeled off by pulling the edge toward the periphery or parallel to the limbus. Few nodules were dissected using a blade. Dissected tissue was sent for histo-pathological examination.

Following removal of the nodules, the underlying corneal surface was smooth. Topical antibiotics were given and a bandage contact lens was placed to induce re-epithelialization. Post-operatively, we started the patient on topical steroid which was tapered over a period of one month to reduce the chances of scarring and recurrence. We also continued the treatment of meibomitis and blepharitis to promote healing and to avoid any foci of infection that would lead to re-occurrence.

Histopathological examination reported that the corneal epithelium overlying the nodule was atrophic and a hyalinized fibrous plaque was present between the epithelium and Bowman's layer (FIG 2). A part of bowman's layer was absent at the right side of the lesion. (H&E, 50x). On Periodic acid Schiff stain, the epithelial basement membrane was thickened and irregular. (Fig 3, Fig 4)

On subsequent follow ups, there was complete clearing of the sub-epithelial opacities and the underlying stroma was also clear.

Written and informed consent was obtained from the patient for the use of photographs for publication of this article.

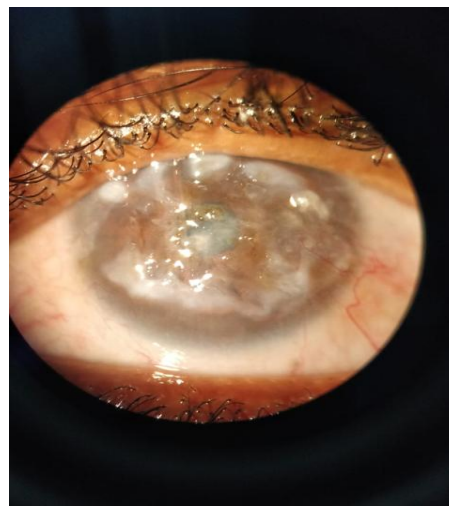


Fig 1



Fig 2



Fig 3

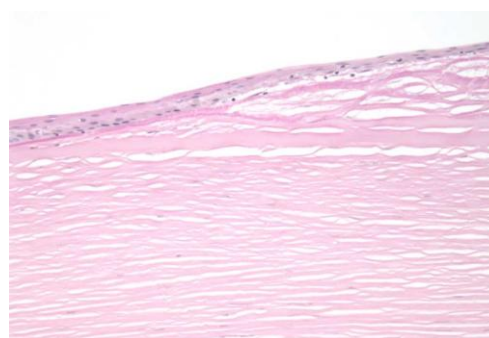


Fig 4

Discussion

Salzmann's nodular degeneration is reported as a bilateral degeneration of cornea by some authors, while other define it as unilateral condition⁵. The incidence is seen more in females than males⁶.

At the scared zone or in the region of the scarring at the cornea, nodules are present which are typically of bluish-grey or-white in colour, elevated above the corneal surface⁵. They may be present at the center or periphery arranged commonly in a circular pattern at the center of the cornea^{7,8}.

The patient are mostly asymptomatic but in acute stage present with pain, redness and photophobia which is due to epithelial break down⁸. The etiology of the degeneration is not clear but chronic inflammatory conditions of the cornea act as predisposing factor⁹⁻¹⁴. A study conducted on of 152 eyes reported some common associations like meibomitis, contact lens use, pterygium, keratoconjunctivitis sicca, peripheral vascularization and exposure keratitis⁹.

Salzmann nodules are present in between a thinned out corneal epithelium and a broken or absent Bowman's layer consisting of a hyper cellular area of extracellular matrix¹⁵. The epithelial cells over the nodules stain negative for matrix metalloproteinase-9 but positive for matrix metalloproteinase-2 (MMP-2)¹⁵. The protein (MMP-2) dissolves type 4 collage which is the main constituent of the basement membrane of the cornea⁴. Hence, the destruction of the bowman's layer may be contributed by these nodules as the overlying cells express the MMP 2 protein⁴. Some hypothesis emphasize the over expression of MMP 2 protein in the basal epithelial cells over the Salzmann nodule, occur by induction due to an elevating nodule¹⁵ However, the exact origin of the nodule is still unclear.

H&E stain typically reports either complete absence or breaks in Bowman's layer and the nodules with very thin over lying epithelium along with irregular arrangement of collagen fibrils present in the sub epithelium.⁴

Light microscopy shows hyalinized dense deposits, collagen fibers arranged irregularly in front of a broken and sometimes absent Bowman's layer^{1,17}. The epithelium in the tissue present over the nodule is attenuated with 2-3 layers of hyper chromatic cells present above 1-2

layers of elongated basal cells which are discontinuous^{1,17}. The corneal stroma contains unevenly distributed keratocytes and disorganized collagen bundles. The unevenly arranged keratocytes in the nodular stroma are mitotically in active, however, their activity is similar to cells involved during corneal repair, the activated fibroblasts or myofibroblasts of the anterior stroma¹.

Surgical removal of the nodules depending upon the clinical picture of the patient along with medical management is the main stay of the treatment as no spontaneous resolution of the nodules has been reported till date⁴. Good visual outcomes have been reported after surgical treatment⁴.

Conclusion

Salzmann's nodular corneal degeneration is a non-inflammatory nodular corneal degeneration which progresses slowly, causing ocular surface discomfort and progressive astigmatism which decreases the best corrected visual acuity^{5,6}

The positive history of diminution of vision which was progressive, increasing astigmatism, association with chronic inflammatory conditions like blephritis and meibomitis and typical findings of nodules on the cornea confirmed by histopathological examination further support our diagnosis – Salzmann's degeneration of cornea in right eye.

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