

A CASE REPORT ON SCHALZMANN NODULAR DEGENERATION

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A 54-year-old woman presented to eye OPD with a 5-year history of painless, progressive loss of vision in both eyes. Gradual, painless loss of vision in both eyes (right greater than left) progressed gradually over last 3 years at both distance and near. She has had 3 updates of her spectacle prescription that have not provided satisfactory vision. There was no history of diurnal variation in vision.

Past Ocular History revealed Myopia, presbyopia, astigmatism. Dry eye syndrome was present with relief of symptoms with artificial tears. Medical History was nonsignificant. No ocular medications. No family history of glaucoma, macular degeneration, blindness or known ocular diseases.

Ocular Exam revealed **best corrected** Visual acuity (VA) with correction in Right eye (OD): 6/9, left eye: 6/6 Intraocular pressure (applanation): OD 16 mm Hg, OS 18 mm Hg. Pupils: Equal, round, 4 mm in dark, 2 mm in light, no relative afferent pupillary defect. Visual fields: Full to confrontation both eyes (OU). Motility: Full OU

Slit Lamp Exam

External: Normal OU. Lids & lashes revealed inspissated meibomian glands with thick waxy secretions and telangiectasia, normal lashes OU.

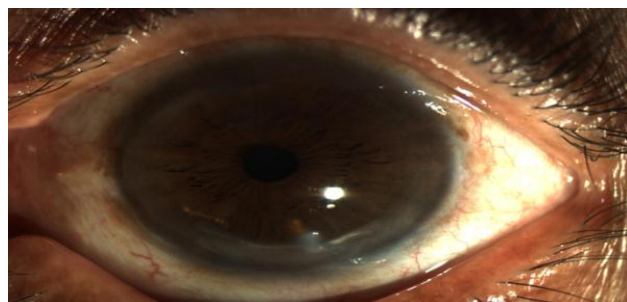
Conjunctiva & sclera: Clear and quiet.

Cornea

OD: Greyish nodules between the corneal epithelium and Bowman's layer in whole cornea extending centrally into the visual axis; iron line at inferior border of the lesion.

OS: Greyish nodules between the corneal epithelium and Bowman's layer in inferonasal region at 5 'o'clock extending centrally into the visual axis.

Anterior chamber: Deep and quiet OU. Iris: Normal architecture OU. Lens: OD: 2-3+ nuclear sclerosis, OS: 1-2+ nuclear sclerosis. Vitreous: Normal OU



Dilated Fundus Exam. Disc: Normal, cup-to-disc ratio 0.3 OU. Vessels: Normal OU. Macula: Normal OU. Periphery: Normal OU.

A diagnosis of Salzmann's nodular degeneration was kept on the basis of the typical clinical image and the clinical history of continuously increasing irregular astigmatism related with worsening of best corrected

visual acuity. She was also diagnosed with nuclear sclerotic cataracts on the right greater than left eye. To improve visual acuity and to decrease the irregular astigmatism a superficial keratectomy was recommended to remove the Salzmann's nodules.

Pre-operative Manifest Refraction		
-2.00 + 5.00 x 20	VA 6/9	OD
-0.75 + 7.50 x 165	VA 6/6	OS

Pathology

Salzmann's nodules are characterized by hyaline deposited anterior to a disrupted or even displaced Bowman's layer, posterior to an atrophic corneal epithelium.^[1]

Salzmann's nodular degeneration (SND) is a infrequent, noninflammatory, gradually progressive degenerative disease of the cornea characterized by the development of nodular bluish grey opacities varying in number and size. It is usually bilateral; most commonly occurring in people aged 50-60 years old, with a female preponderance; and often associated with a history of antecedent corneal inflammation. The clinical features usually depend on the location of the nodules. Generally, the nodules of SND are bluish white to gray in color, 1-2 mm in size, and round, conical or prismatic in shape. The overlying Bowman's layer is usually absent from the nodular areas and is partially replaced by granular Periodic Acid Schiff-positive eosinophilic material resembling the basement membrane. Diagnostic investigations include ultrasonic pachymetry, anterior segment optical coherence tomography, ultrasound biomicroscopy, and confocal microscopy. The majority of patients respond well to conservative management with topical lubricants; severe cases may require surgical intervention. The various surgical modalities described include superficial keratectomy, which may be combined with phototherapeutic keratectomy and keratoplasty. Various modifications of these procedures include the use of alcohol-assisted epithelial delamination, intraoperative mitomycin-C or amniotic membrane transplantation to make the procedure easy, reduce the risk of recurrence and improve postoperative comfort. Recurrences are rarely reported; overall, the visual prognosis following treatment is optimal.^[7]

DISCUSSION

Salzmann's nodular corneal degeneration was described by Katz in 1930 following a 1925 case series on the condition published by Maximilian Salzmann, although Salzmann concedes that a case published by Ernst Fuchs in 1901 appears to be the first published case of this corneal degeneration.^[1]

Salzmann's nodular corneal degeneration is characterized by bilateral gray-white elevated nodules anterior to Bowman's layer, which may be visually significant, cause foreign body sensation, or be asymptomatic. It is most common in middle-aged

women. The cause of this degeneration is unknown; however, it has been associated with chronic ocular surface inflammation.^[1-6] A case series of 152 eyes concluded that the most common associations were, in descending order, meibomian gland dysfunction (MGD), contact lens wear (especially hard contact lenses), peripheral vascularization, pterygium, keratoconjunctivitis sicca, and exposure keratitis.^[1] Another case series of 180 eyes also identified MGD as the most common comorbidity.^[2]

Conservative treatment consists of management of the underlying etiology, such as MGD. Eyelid hygiene and doxycycline may be considered to treat the MGD prior to surgical treatment. Contact lens cessation or re-fitting may also be beneficial. Medical management is successful in preventing the need for surgery in most cases; the indications for surgery are (1) discomfort; (2) reduced vision due to progressive increase in astigmatic error. Surgery is uniformly successful in removal of the nodule and almost invariably successful in reducing the astigmatic error in allowing for improvement in best corrected visual acuity, usually with a much weaker spectacle prescription. Stabilization of the refractive error usually takes 3 to 6 weeks. In rare cases of neglected nodules within the visual axis, anterior stromal haze may compromise the final visual results.

Whereas subtle recurrences are common in most cases over a 5 to 15 year period, visually significant recurrences are uncommon (5 to 20%) and can be minimized with meticulous control of the etiology responsible for the condition.^[1,2] Those cases can be treated with repeat superficial keratectomy. If anterior stromal haze persists in the visual axis after treatment of primary or recurrent nodules, phototherapeutic keratectomy can be offered.^[1]

Superficial keratectomy of astigmatism-inducing Salzmann's nodules should always be performed prior to cataract surgery. Stable keratometry readings and IOL calculations cannot be reliably obtained until 3 to 6 weeks after surgery. Toric IOLs should be avoided because of the risk of recurrent nodules and induction of post-operative astigmatism.

Since the diagnosis of Salzmann's nodular corneal degeneration is clinical.

CONCLUSION

Salzmann's nodular corneal degeneration is a non-inflammatory, slowly progressive nodular corneal degeneration that may induce ocular surface discomfort and progressive astigmatism with decreased best corrected visual acuity. The diagnosis is based on clinical examination. Topography is helpful in evaluating the contribution of the nodule to visual impairment. Superficial keratectomy is a successful treatment option, and visually significant recurrence afterward is relatively rare.

Differential diagnosis^[4]

- Corneal scarring
- Spheroidal degeneration

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