Contact Lens & Anterior Eye 34 (2011) 173–178

Contents lists available at ScienceDirect

Contact Lens & Anterior Eye

journal homepage: www.elsevier.com/locate/clae

Salzmann's nodular corneal degeneration (SNCD): Clinical findings, risk factors, prognosis and the role of previous contact lens wear

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ARTICLE INFO

Keywords:
Salzmann's
Nodular
Degeneration
Soft contact lens
Prognosis
Risk factors

ABSTRACT

Purpose: To revisit the clinical presentations of Salzmann's nodular corneal degeneration (SNCD) and to identify risk factors, occurrence and prognosis, and in particular to assess the role of previous contact lens wear as an aetiological factor.
Methods: Retrospective case note review of all cases of Salzmann's nodular degeneration over the last twenty years. We analysed epidemiological features, characteristics of lesions, risk factors and final outcomes.
Results: Thirty eyes (19 patients) were identified with SCND. Eleven patients had bilateral disease. Our cohort included 14 female (73.7%) and 5 males (26.3%). Average age at presentation was 58.9 (range 30–75) years. Follow up range was 0–13 years. The most common presenting symptom was foreign body sensation (68.4%). Ocular pathologies were: dry eyes (56%), chronic blepharitis (32%), trichiasis (8%), trachoma (32%), previous ocular trauma (8%), and previous ocular surgery (21.4%). Sixteen percent of cases were soft monthly disposable contact lens wearers. None of our patients with rigid contact lens wear developed Salzmann's nodules. Surgical excision was needed in 4 cases (13.3%). Two of them developed recurrent disease.
Conclusion: Salzmann's nodular corneal degeneration is a disorder affecting middle-aged white women predominantly, and seems to be associated with concomitant chronic MGD, dry ocular surface, soft contact lens wear and previous ocular surgery. The prognosis is very good, and most patients are "successfully" treated with medical management alone, and therefore correct diagnosis of the disease is paramount. If indicated, various surgical options are available and give good outcomes. However, Salzmann's nodules can recur after penetrating keratoplasty.

1. Introduction

Salzmann's nodular corneal degeneration (SNCD) is a worldwide occurring, relatively rare, non-inflammatory, slowly progressive disease originally described by Maximilian Salzmann in 1925 [1]. However, Ernst Fuchs may have made the initial case observation in 1901 [2].

SNCD is classically described as bluish-greyish nodular corneal opacities of various sizes (mostly 1–3 mm) occurring in isolation or in multiple numbers anywhere on the cornea (Fig. 1(a–b)). Salzmann's nodules can occasionally take up fluorescein staining (Fig. 2). Histopathological findings involve thinning or denudation of the epithelium, destruction of Bowman's layer, duplication of the epithelial basement membrane and disorganisation of collagen lamellae in the superficial anterior stroma. These histological findings are entirely non-specific and can be seen in scarring from any cause. The cause of the degeneration is uncertain and still debated upon [3], but associations with chronic ocular irritation and inflammation have been made. It may be idiopathic; however it is often associated with pre-existing corneal disease. SNCD has been associated with trachoma, interstitial keratitis, vernal keratoconjunctivitis, phlyctenular keratoconjunctivitis, ocular trauma, corneal exposure, measles, scarlet fever, and previous ocular surgery. The purpose of this study is to revisit the clinical presentations of SNCD and to identify risk factors, occurrence and prognosis. In particular, the role of previous contact lens wear in the development of SNCD will be investigated.

2. Patients and methods

Retrospective case note review of all cases of Salzmann's nodular degeneration that we have identified in our centre (Birmingham and Midland Eye Centre) over the last twenty years (March 1990–April 2010). We analysed epidemiological features, characteristics of lesions, risk factors and final outcomes.

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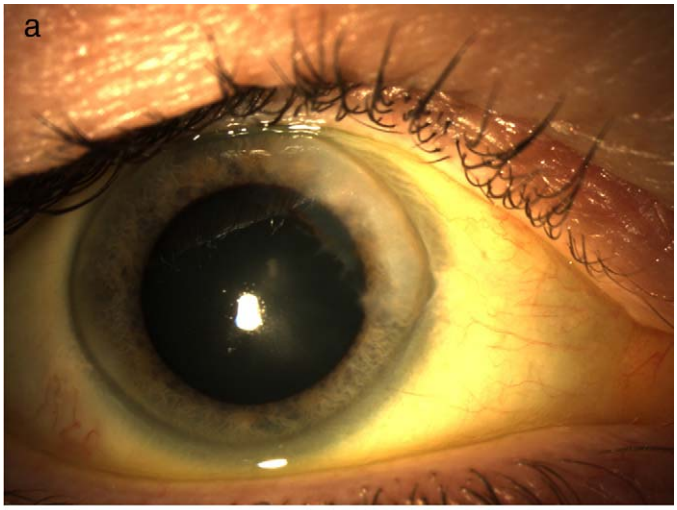
doi:10.1016/j.clae.2011.02.004



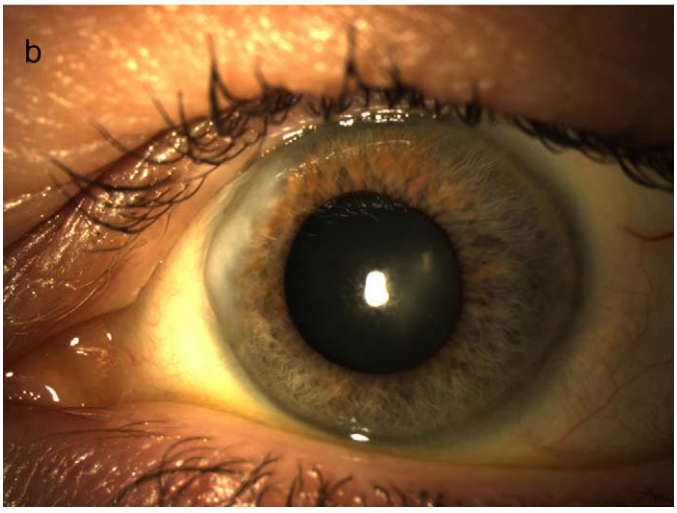


Fig. 1. (a–b) Salzmann's nodules appear as bluish-white nodules raised above the surface of the cornea. It can be single, multiple (separated or confluent).

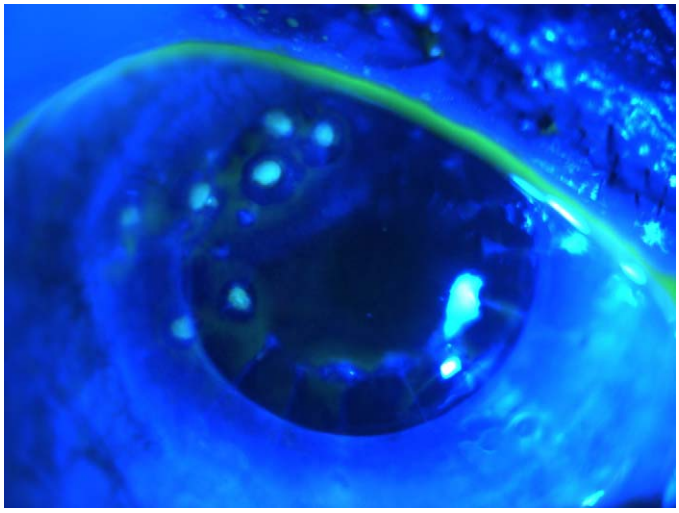


Fig. 2. Cobalt blue light shows fluorescence staining of Salzmann's nodules.

3. Results

Thirty eyes (19 patients) were identified with the clinical diagnosis of Salzmann's nodular degeneration. Our cohort included 14 females (73.7%) and 5 males (26.3%). Average age at presentation was 58.9 (range 30–75) years. Follow up period varied according to initial presentation, severity, and progression of the disease; and ranged from 0 to13 years. More than half of patients were white Caucasian (11/19). The remaining were Asian (7/19) and Afro-Caribbean (1/19). Demographical data are summarised and compared with other studies in Table 1.

Eleven patients had bilateral disease. Salzmann's nodule distribution varied among patients. All eyes had peripheral corneal nodules. 10% of all eyes had lesions in the central cornea involving visual axis. We looked at the distribution of corneal lesions, which were as follows: super-nasal quadrant 28.2%, super-temporal quadrant 23%, infra-nasal quadrant 20.5%, infra-temporal quadrant 17.9%, and central cornea 10.3%. All of which were not statistically significant (Table 1). Central corneal nodules were seen in the eyes of Asian females who also showed signs of ocular surface trachoma. These females had both eyes affected with SCND but only one eye manifested Salzmann nodules in the central cornea.

Peripheral corneal vascularisation (360°) was seen in one patient (two eyes) who had bilateral disease and a history of bilateral penetrating keratoplasties. Penetrating keratoplasties were performed to treat severe corneal scarring from herpes simplex keratitis.

The majority of patients (68.4%) complained of foreign body sensation. Other symptoms included "white spots on cornea" (5.3%) and reduced vision (5.3%). However, 15.8% were asymptomatic and referred by their optometrist to exclude the presence of treatable corneal disease (Table 1).

We looked at other ocular pathologies that were documented at the time of first presentation and these included: dry eyes (56%), chronic blepharitis (32%), trichiasis (8%), trachoma (32%), previous ocular trauma (8%), previous ocular surgery (21.4%) of which 8% were post pterygium surgery (Table 1). We identified that 16% of cases were soft monthly disposable contact lens wearers but they had no other ocular pathology. None of our patients with rigid contact lens wear developed Salzmann's nodules.

Management of SNCD involved: ocular lubrication (66%), topical anti-inflammatory (33%) mainly Prednisolone 0.5% eye drops, and surgical excision in 4 cases (13.3%). Two underwent penetrating keratoplasties, one had simple surgical excision, and one had anterior lamellar keratoplasty. The disease has recurred in two patients (2 eyes), both had penetrating keratoplasty. Finally, we reported final visual outcomes i.e. visual acuities measured at last hospital visit: 21 eyes $\geq$ 6/9, 1 eye = 6/12, and 4 eyes $\leq$ 6/36 (all trachoma cases). No visual acuities data at last visit for 4 eyes.

4. Discussion

Originally Salzmann described 23 patients with bluish-white corneal lesions, 18 of whom were females [1]. Later studies by Farjo et al. and Graue-Hernandez et al. [2,3] supported Salzmann observations that SNCD affect mainly middle aged females. Our cohort reflected the same trend; 73.7% of our patients were females and the average age was 58.9 years (30–75).

Furthermore, our study showed a preponderance of SNCD to affect the white population (57.9%). Graue-Hernandez reported the majority of his cases were white (76.1%) in a group where the total white population was only 47.8% [3]. However he suggested that access to medical care may have been more readily available to certain populations, which can introduce bias in the study sample. Therefore, more studies are needed to support these findings.

As the majority of the patients in our study were middle aged females and older males, and the commonest associations with SNCD were dry ocular surface (56%), chronic meibomian gland dysfunction (MGD) 32%, it can be postulated that there may be an