Original Research Article

Corneal degenerations in central India: Clinical profile and visual impact

Garima Singh¹*, Kavita Kumar¹, Sooraj Singh Kubrey¹

¹Dept. of Ophthalmology, Gandhi Medical College, Bhopal, Madhya Pradesh, India

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A B S T R A C T

Aims: To study cases of corneal degenerations presenting to a tertiary eye care centre in central India with regards to their clinical profile, socio-demographic parameters, association with dry eye and their visual impairment at presentation.

Materials and Methods: Cases of corneal degenerations presenting to our Cornea Clinic in a span of one year were identified and categorized. Details were noted regarding age, sex, socioeconomic status, residential locality, occupation, working conditions and prior ocular history. Dry eye assessment was done using Schirmer test and patients were sorted under various categories of visual disability.

Results: We came across a total of 127 cases of corneal degeneration, among which the maximum number (74.80%) were of Arcus Senilis (95 cases), followed by 10.24% Spheroidal Degeneration (13), 7.09% Band Shaped Keratopathy (9), 5.51% Crocodile Shagreen (7), 1.57% Salzmann Nodular Degeneration (2) and 0.79% Terriens Marginal Degeneration (1 case). The mean age was 61.66 years and the male: female ratio was 1.89:1. Most cases belonged to low socioeconomic status (48.03%), urban locality (65.35%) and were involved in indoor work (67.72%), with the exception of spheroidal degeneration. Only 12.60% cases of degeneration had Schirmer 1 values suggestive of dry eye problem (<10mm wetting). Majority of the cases (32.28%) fell in Category 2 of visual disability (75% impairment), excluding the 33.86% of cases which had even <20% impairment, and were constituted mainly by arcus senilis.

Conclusion: The study sheds light on the profile of cases of corneal degenerations, some findings in accordance with the existing data and some not, in particular relevance to the current trend and the specified region.

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1. Introduction

In India, the burden of corneal blindness is said to be immense. It is estimated that there are approximately 6.8 million people who have vision less than 6/60 in at least one eye due to corneal diseases (according to a report by National Programme for Control of Blindness); of these, about a million have bilateral involvement.¹,² It is expected that the number of individuals with unilateral corneal blindness will increase to 10.6 million by 2020.² There are currently 120,000 corneal blind persons in the country, with an estimated addition of 25,000-30,000 corneal blindness cases every year.¹ Corneal degenerations are one of the important causes of loss of corneal transparency which, apart from causing diminution or blurring of vision, can cause multiple other ocular problems. They are basically secondary disorders that usually occur after previous specific insult, result from a specific disease or simply from aging. Changes caused by inflammation, maturity or systemic disease result in deposition, thinning or vascularization of the corneal tissue. Some degenerations do not require any active intervention, only symptomatic treatment can provide relief, while others require active interventions like excimer laser keratectomy or lamellar or penetrating keratoplasty. But in majority of cases, the long term prognosis may not be so good, because the disease is progressive in nature or recurrence can occur after surgery. In some degenerations, treating the primary disease can be
helpful. With these problems in view, the present study was conducted to evaluate the clinical and socio-demographic profile of cases of corneal degeneration in this belt of Central India, so that early detection and intervention could be possible.

2. Material and Methods

This was a hospital based cross-sectional study performed on patients attending the Cornea Clinic in a tertiary eye care centre in Central India, within a time span of one year.

All cases of corneal degenerations were recorded under the headings of age, sex, address, locality – urban/rural, socioeconomic status (as per Kuppuswamy scale), occupation and working conditions. The history was taken in detail as to the symptomatology with its onset, duration and progression of symptoms. Points also noted were family history, past history, systemic illness, prior ocular disease, trauma / surgery or any treatment undertaken. Best corrected visual acuity on Snellen's was noted and all patients were categorized under various categories of visual disability and impairment in accordance with the guidelines given by the Ministry of Social Justice and Empowerment, Government of India (2001).

Thorough anterior segment examination was done on slit lamp and various degenerations were identified and classified clinically. Schirmer test was done to evaluate dry eye disorders, which included both parts - Schirmer 1 which gave the amount of total secretion, and Schirmer basal test, after anaesthetizing the conjunctival sac with 4% xylocaine, which gave the reading for the amount of basal secretion. However for categorizing patients under dry eye, Schirmer 1 values were used due to lack of standard accepted cutoff values for Schirmer with anaesthesia. Fundus examination and B Scan was done to rule out any posterior segment abnormality.

Patients with infectious pathology, posterior segment abnormalities, and those unwilling for participation, were excluded from the study. Data was collected, tabulated and analysed with the appropriate tests where applicable.

3. Observations

In our study, we came across a total of 127 cases of corneal degeneration, among which the maximum number (74.80%) were of Arcus Senilis (95 cases), followed by 10.24% Spheroidal Degeneration (13), 7.09% Band Shaped Keratopathy (nine), 5.51% Crocodile Shagreen (seven), 1.57% Salzmann Nodular Degeneration (two) and 0.79% Terriens Marginal Degeneration (one case). It was further noted that overall 87.40% cases were bilateral, and 12.60% were unilateral, with all cases of arcus senilis, crocodile shagreen and terriens marginal degeneration being bilateral.

The mean age of degenerations in this study was 61.66 years, with about 41.73% of cases being in the age group 60-70 years. [Table 2] shows the further distribution of the cases according to age. We found that males comprised 65.35% of cases of corneal degeneration, and females 34.64%. Overall, the maximum proportion of males was seen in spheroidal degeneration (84.62%). Both the cases of Salzmann nodular degeneration were female, and the only case of Terriens marginal degeneration was male.

It was observed that 48.03% cases belonged to low socioeconomic status, 43.31% to middle socioeconomic status and 8.66% cases to high socioeconomic status. In terms of residential status we found that overall 65.35% cases belonged to urban locality, while 34.65% belonged to rural area. However in case of spheroidal degeneration, majority (53.85%) belonged to rural area.

On analysing all the cases in terms of their occupation, we found that 67.72% patients were employed in indoor work, while 32.28% had outdoor activities. However an exception was spheroidal degeneration where 61.54% had outdoor work and 38.46% indoor work. In a few cases we also noted history of some kind of previous ocular condition. 14.96% cases of degeneration had a prior history of trauma, 8.66% had history of some kind of keratoconjunctivitis/ulcer, and 31.50% of previous ocular (cataract) surgery. [Table 3]

On evaluating the cases for dry eye, on the basis of Schirmer 1 values, we found 16 patients (12.60%) to be having Schirmer 1 values suggestive of dry eye problem; with two patients having Schirmer 1 \(<=5\)mm and 14 having values between 5-10mm. Rest 111 patients of degeneration (87.40%) had a sufficient wetting of Schirmer >10 mm. [Table 4]

In this study we categorised all the cases under different categories of visual impairment based on Government of India’s disability guidelines. Among corneal degenerations we found the following distribution – maximum fell in Category 2 (75% impairment) – 32.28% cases, followed by Category 1 (40% impairment) – 14.17%, Category 0 (20% impairment) – 11.02% cases, Category 3 (100% impairment) – 3.94%, Category 4 (100% impairment) – 1.57%. One eyed person category (30% impairment) included 3.15% cases. However a large group of patients fell in the category of <20% impairment, where the BCVA of both eyes was good enough such that they did not even reach the 20% impairment (category 0). They comprised 33.86% of all degenerations, with arcus senilis cases forming 95.35% of this group. Among arcus senilis patients too, the maximum (43.16%) fell in this category only. [Table 5] elaborates on the same.

4. Discussion

According to Waring GO et al (1998) \(^5\) the distribution of corneal degenerations is heavily influenced by two major factors – geographic location and age. They studied 105 consecutive eyes of patients having corneal degenerations
Table 1: Categories of visual disability (as per Best corrected visual acuity – BCVA)

| Category | Better eye | Worse eye | % Age Impairment |
|----------|------------|-----------|------------------|
| Category 0 | 6/9-6/18 | 6/24-6/36 | 20% |
| Category 1 | 6/18-6/36 | 6/60 to Nil | 40% |
| Category 2 | 6/40-4/60 or field of vision 10-20\(^0\) | 3/60 to Nil | 75% |
| Category 3 | 3/60-1/60 or field of vision 10\(^0\) | Finger count at 1 ft. to Nil | 100% |
| Category 4 | Finger count at 1 ft. to Nil or field of vision 10\(^0\) | Finger count at 1 ft. to Nil | 100% |
| One eyed persons | 6/6 | Finger count at 1 ft. to Nil or field of vision 10\(^0\) | 30% |

Table 2: Distribution of cases of corneal degenerations according to age

| S. No | Degeneration | <= 20 yrs. | 20-40 yrs. | 40-60 yrs. | >70 yrs. |
|-------|--------------|------------|------------|------------|---------|
| 1     | Spheroidal Degeneration | - | 2 | 3 | 8 |
| 2     | Band Shaped Keratopathy | 4 | 1 | 2 | 2 |
| 3     | Crocodile Shagreen | - | - | - | 7 |
| 4     | Salzmann Nodular Degeneration | - | - | - | 2 |
| 5     | Terriens Marginal Degeneration | - | - | - | 1 |
| 6     | Arcus Senilis | - | - | 35 | 60 |
| Total | 4 (3.14%) | 3 (2.36%) | 40 (31.50%) | 78 (61.42%) |

Table 3: Association of various degenerations with previous ocular conditions

| S. No | Degeneration | H/O trauma/ FB entry | H/O ulcer / keratoconjunctivitis | H/O ocular surgery |
|-------|--------------|----------------------|----------------------------------|--------------------|
| 1     | Spheroidal Degeneration | 5 | 7 | 1 |
| 2     | Band Shaped Keratopathy | 4 | 1 | 1 |
| 3     | Crocodile Shagreen | - | - | 2 |
| 4     | Salzmann Nodular Degeneration | - | 1 | 1 |
| 5     | Terriens Marginal Degeneration | - | - | 1 |
| 6     | Arcus Senilis | 10 | 2 | 34 |
| Total | 19 (14.96%) | 11 (8.66%) | 40 (31.50%) |

Table 4: Association of various degenerations with dry eye (Schirmer 1)

| S. No | Degeneration | <= 5 mm | 5-10 mm | >10 mm |
|-------|--------------|----------|---------|--------|
| 1     | Spheroidal Degeneration | 1 | 2 | 10 |
| 2     | Band Shaped Keratopathy | 1 | 1 | 7 |
| 3     | Crocodile Shagreen | - | 1 | 6 |
| 4     | Salzmann Nodular Degeneration | - | - | 2 |
| 5     | Terriens Marginal Degeneration | - | - | 1 |
| 6     | Arcus Senilis | - | 10 | 85 |
| Total | 16 (12.60%) | 111 (87.40%) |

and they found – corneal arcus (70.5%), crocodile shagreen (14.3%), spheroidal degeneration (9.5%), calcific band keratopathy (7.6%) and Salzmann nodular degeneration (4.8%), among others. The findings in our study were in accordance with these observations, with few differences which could be due to geographical variation.

Krachmer (2011)\(^6\) mentions that corneal degenerations may be unilateral or bilateral and are often asymmetric. Hashemi H et al (2014)\(^7\) also noted in their study on arcus senilis, that 98.4% cases were bilateral. Similar results regarding crocodile shagreen were observed by Ansons AM et al (1989)\(^8\), who found such corneal mosaic patterns to be almost universally bilateral. Similar observations were noted in by us.

In our study, maximum number of patients of corneal degeneration were in the age group 60-70 years (41.73% cases) and the mean age of presentation was 61.66 years. According to Waring GO et al (1998),\(^5\) in general the older the age of population, the more prevalent the corneal degeneration, especially those normally associated with
ageing. In multiple other studies also, the prevalence of corneal arcus was seen to increase with age; a fact reaffirmed by Cooke NT (1981)9 Vurgese S et al10 in 2011 and Hashemi H et al9 in 2014. Our findings reflect the same except for a dip in the number of cases in the >70 yrs age group, which could be explained by the lesser life span of Indians (esp. in rural setup) and the reluctance of people in the >70 yrs age group to visit a hospital for such minor ocular ailments.

In the present study, all the degenerations had an increasing prevalence with age, with the exception of band shaped keratopathy, which had 55.56% cases in the <30 years age group. This could be explained by the fact that apart from age, other factors are also instrumental in the development of band keratopathy like chronic intraocular inflammation, uveitis, trauma etc. Similar inference was noted in a study done by Yang P et al11 in 2011, where they concluded that band shaped keratopathy mostly occurs in relatively younger patients with a long course of chronic intraocular inflammation.

In our study the male: female ratio for corneal degenerations was 1.89:1, with males comprising 65.35% of overall cases. However the maximum proportion of males was seen in spheroidal degeneration (84.62%), which could be attributed to the occupational exposure difference between men and women. Wu R et al (2010),12 in his study on risk factors of arcus senilis found that male gender was a significant risk factor for arcus. Similarly, Hashemi H et al (2014 )13 also found that the prevalence of arcus was significantly higher in men (p<0.001). Even though in our study we found that 66.32% of patients of arcus senilis were male, still male gender was not found to be significantly associated with it (p-value=0.69481). An exception was however seen with Salzmann nodular degeneration, where both the cases in our study were females. Salzmann (1925)14 in his original description of the disease, reported that among 23 patients, 18 were females. Two of the largest case series on Salzmann nodular degeneration also reported a female preponderance: Farjo et al14 in 2006 (89.2% females) and Graue-Hernandez et al15 in 2010 (72.2% females).

In our study, we found that 48.03% cases belonged to low socioeconomic status, 43.31% to middle socioeconomic status and 8.66% cases to high socioeconomic status. These observations mostly represent the pattern of patients coming to our institute. We did not find any previous study reporting the relationship of socioeconomic status with corneal degeneration. The slightly more prevalence of degeneration in low socioeconomic group could be due to low awareness and education, or even delay in treatment of pre-existing ocular conditions.

Regarding demographic distribution, we found that overall 65.35% cases belonged to urban locality, while 34.65% belonged to rural area. However in case of spheroidal degeneration, majority (53.85%) belonged to rural area, and 61.54% were involved in outdoor work. Such a scenario could be attributed to the fact that they have more exposure to ultraviolet radiation and micro-trauma due to sand, dust, wind etc. which are known to be directly implicated in spheroidal degenerations (Krachmer 2011, Norm M 1991, Urrets-Zavalia et al 2007),6,16,17

In our study we found that a few cases did have a significant past history of either ocular trauma, surgery, keratoconjunctivitis/ ulcer or medical treatment for some ocular condition. It was especially high in cases of spheroidal degeneration (92.31%) and band keratopathy (55.56%). According to Krachmer (2011),6 secondary spheroidal degeneration occurs in diseased eyes or cornea associated with other ocular pathology. Similarly for band shaped keratopathy also, general causes stated are chronic ocular disease, uveitis, chemicals (eye drops, irritants) etc.

Not much association has been found out of corneal degenerations with dry eye, with the exception of perhaps band shaped keratopathy and salzmann nodular degeneration (Lemp MA 1977, Krachmer 2011).6,18 Regarding Salzmann degeneration it has been postulated that androgen deficiency may lead to meibomian gland dysfunction, evaporative dry eye and reduced tear production with poor ocular surface as well as subsequent formation of nodules. Farjo AA et al (2006)14 and Graue-Hernandez et al (2010)15
found Meibomian gland disease to be present in 54.8% and 41.7% of cases of Salzmann respectively. Hamada S et al (2011), in his study on risk factors of Salzmann nodular degeneration, found dry eye to be present in 56% of cases. In our study however, both the cases of Salzmann had Schirmer 1>10mm with no evidence of dry eye.

In our study we categorised all the cases under different categories of visual impairment based on Government of India’s disability guidelines. We observed that corneal degeneration cases which have a predominant central corneal involvement, like spheroidal degeneration and band shaped keratopathy, had a marked more visual impairment at presentation than those degenerations which affect peripheral cornea more, like terriens marginal degeneration and peripheral crocodile shagreen. This was also noted in cases of arcus senilis, where only the peripheral cornea is involved, and the majority (43.16% cases) had visual impairment even less than 20%. However the final and accurate assessment of visual impairment could not be made because of varying lens status in all patients (cataract of different grades, pseudophakia, posterior capsule opacification). We were also limited by the fact that there have not been any previous studies to compare which used similar disability guidelines and categories.

5. Conclusion
The study sheds light on the profile of cases of corneal degenerations, some findings in accordance with the existing data and some not, in particular relevance to the current trend and the specified region. All these cases if diagnosed at an early stage and treated by appropriate medical or surgical techniques can prevent progression and subsequent visual morbidity. Emphasis on health education and awareness in general population will definitely decrease the impairment due to these degenerative conditions. Special attention must be paid to those group of people more prone to certain degenerations like people having more outdoor work and exposure to sunlight, trauma, and those having prior ocular diseases.

6. Prior Publication
None.

7. Source of Funding
None.

8. Conflict of Interest
None.

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Author biography
Garima Singh Post Graduate
Kavita Kumar Professor and HOD
Soooraj Singh Kubrey Associate Professor

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