Prevalence of spheroidal degeneration of cornea and its association with other eye diseases in tribes of Western Rajasthan

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Purpose: To determine the prevalence of spheroidal degeneration of cornea (SDC) and its association with other eye diseases in the tribes of South-West Rajasthan. **Methods:** A total of 5012 patients were examined on slit lamp for the diagnosis of SDC. Diagnosis of SDC was made based on presence of amber granules in the superficial stroma of peripheral interpalpebral cornea with increasing opacification, coalescence and central spread or nodular and hazy surrounding stroma and divided in three stages. **Results:** The prevalence of SDC was 10.7%. Around 55% of the total of 535 cases examined were found to have Stage I followed by Stage II (32%) and Stage III (13%). The prevalence is greatest in both men and women over 70 years of age. The severity of SDC is greater in men. SDC was significantly associated with pterygium and pseudocapsular exfoliation. **Conclusion:** Extreme temperature, low humidity, dust, high wind, and microtrauma caused by sand particles are the probable etiologies for higher prevalence of this kind of degeneration in this region.

Key words: Arcus senilis, cortical and nuclear cataract, pseudocapsular exfoliation, pterygium, spheroidal degeneration of cornea

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Spheroidal degeneration of cornea (SDC) is also called Labrador keratopathy, Bietti nodular hyaline band-shaped keratopathy, climatic droplet keratopathy, proteinaceous corneal degeneration, elastotic degeneration, Fisherman's keratopathy, and Eskimo's corneal degeneration. SDC was first described by Bietti in 1955. It is a common degeneration of the cornea which is usually bilateral and interpalpebral. It is more common in males and involves the cornea and conjunctiva, and is characterized by translucent, golden brown, spheroid-like deposits in the superficial stroma.

Fraunfelder *et al*. divided this condition into three basic forms.[1,2]

1. The primary corneal type associated with increasing age
2. The secondary corneal type associated with significant preexisting pathology; and
3. The conjunctival type.

All cases consist of extracellular proteinaceous deposits with elastotic degeneration, which are thought to be secondary to the combined effects of genetic predisposition, actinic exposure, age, and perhaps various kinds of environmental trauma other than sunlight, such as wind. The pattern is similar to that of other ultraviolet (UV) light-associated degenerations, such as pinguecula. The composition is not lipid despite its "oil droplet" appearance.[3]

The degeneration is a common cause of blindness in countries with climatic extremes.[4] It is common in areas with high levels of exposure to UV radiation, including the Labrador, Saudi Arabia, the Dahlak Islands, India, South Africa, and other Arctic and tropical areas in which the population is exposed to high levels of direct and/or reflected sunlight. The higher prevalence in these parts of the world suggests that UV light exposure plays a causative role in the etiology of SDC.

Clinically, SDC is classified into three stages.[5,6]

Stage 1: Spheroids confined to the corneal periphery. Vision is not affected by the degeneration. Stage 2: Spheroids extending to interpalpebral fissure. Vision is affected and may be reduced to 6/60. Stage 3: Same as stage 2 with addition of nodules or excavated areas. Vision is reduced to counting fingers or hand movements.

Association with other ocular diseases such as glaucoma, pseudocapsular exfoliation, pterygium, trachoma, and arcus senilis has been reported. Association with malnutrition has also been suggested.[6]

Prevalence of SDC is high among peoples exposed to direct sunlight, particularly in regions of high UV radiation and among those peoples exposed to micro trauma (such as in a desert). Prevalence increases with age and is higher among males compared to females.[5,6]

The aim of this study is to determine the prevalence of SDC in the region of South-West Rajasthan and determine the association of SDC with other ocular diseases.

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Methods

This is a hospital-based cross-sectional study. The study group consisted of all patients attending the outpatient and inpatient departments of base hospital, vision centers, and eye camps. Only those patients who were residents of Pali, Jalore, Sirohi, and Udaipur districts of Rajasthan (South-West Rajasthan) were included. A total of 5012 (2538 male and 2474 female) patients were selected for the study. Written consent of all the patients was taken.

All patients were examined on slit lamp. Diagnosis of SDC was made based on the following findings.[6]

- Amber granules in the superficial stroma of peripheral interpalpebral cornea
- Increasing opacification, coalescence, and central spread
- Advanced lesions – nodular and surrounding stroma hazy.

Degeneration was divided into three stages.[6]

The presence of pterygium and arcus senilis was confirmed during the same examination. Diagnosis of cataract and its grading was done on slit lamp with pupil fully dilated. Pseudocapsular exfoliation was diagnosed by typical granular deposits seen on the anterior lens surface by slit-lamp examination with pupil fully dilated.

Glaucoma (primary open-angle glaucoma [POAG]) was diagnosed on the basis of disc and field examination (ophthalmoscopic and Humphrey field analyzer, respectively), repeated tonometry, and gonioscopy. A cupped disc, field loss, ocular pressure above 21 mmHg, and an open angle were the criteria for open-angle glaucoma.

The Institutional Ethics Committee on Human Subjects Research, 2013–2014, granted approval, subsequent to which data collection for the study was initiated. Descriptive statistical analysis has been carried out in the present study and results on categorical measurements are presented in a number (%). The relative risk (RR), its standard error, and 95% confidence interval (CI) have been calculated according to Altman, 1991.

The RR or risk ratio is given by:

$$RR = \frac{a/(a+b)}{c/(c+d)}$$

With the standard error of the log *RR* being,

$$\mathrm{SE}\{\mathrm{In}(RR)\} = \sqrt{\frac{1}{a} + \frac{1}{c} - \frac{1}{a+b} - \frac{1}{c+d}}$$

and 95% CI

$$95\%\,\mathrm{CI} = \mathrm{Exp}(\mathrm{In}(RR) - 1.96 \times \mathrm{SE}\{\mathrm{In}(RR)\}) \text{ to } \mathrm{Exp}(\mathrm{In}(RR) + 1.96 \times \mathrm{SE}\{\mathrm{In}(RR)\})$$

Where zeros cause problems with computation of the RR or its standard error, 0.5 is added to all cells (a, b, c, d). $P < 0.05$ was taken as statistically significant. The statistical software, statistical product and service solutions (SPSS for Windows, Version 16.0, Chicago, SPSS Inc.) was used for the analysis of the data, and Microsoft Word and Excel have been used to generate graphs, tables, etc.

Results

To calculate the prevalence rate for SDC, only one eye was taken into consideration in bilateral involvement. The figures presented refer only to eye with SDC of lesser grade. This is done to avoid overestimation. Presence of associated ocular diseases even in one eye was considered as positive association. Among 5012 patients (2538 males and 2474 females), 535 were found to have SDC prevalence of 10.674%.

Total male SDC cases were 301 (prevalence – 11.86%). Total female SDC cases were 234 (prevalence – 9.458%). Prevalence of SDC was higher among males. The distribution of SDC cases in accordance with age and sex is shown in Table 1 and Figs. 1 and 2.

Table 2 represents RR between male and female population among different age groups. In both men and women, the prevalence increased significantly with increasing age. The prevalence in both male and female patients is greatest in those over 70 years of age.

The prevalence in males was higher than females in all age groups, the greatest difference being in the age group of 40–50 years ($P < 0.001$).

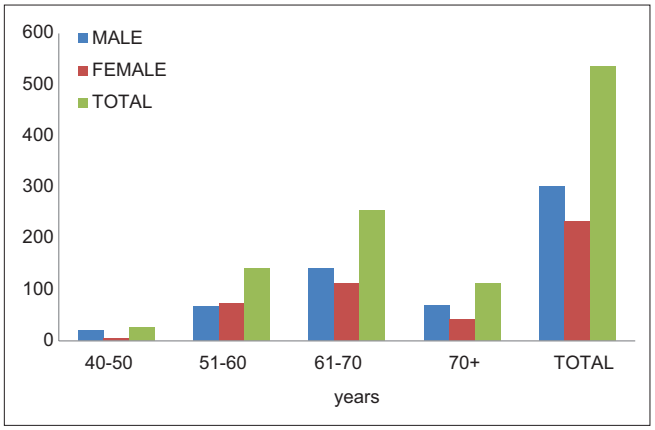

Figure 1: Distribution of spheroidal degeneration of cornea cases in accordance to age and sex

Table 1: Distribution of spheroidal degeneration of cornea cases in accordance to age and sex

Age (years)	Male				Female			
	Stage I	Stage II	Stage III	Total	Stage I	Stage II	Stage III	Total
40-50	17	3	1	21	2	2	1	5
51-60	37	24	7	68	38	25	11	74
61-70	77	47	18	142	64	36	12	112
70+	39	23	8	70	21	12	10	43
Total	170	97	34	301	125	75	34	234

Table 2: Relative risk between males and females

Age (years)	Males		Females		Relative risk	Significance (level of data between sexes)
	Number with SDC (%)	Total number of investigated	Number with SDC (%)	Total number of investigated		
40-50	21 (8.93617)	235	5 (2.212389)	226	1.641804	<0.001
51-60	68 (7.934656)	857	74 (7.51269)	985	1.031793	0.7319
61-70	142 (12.76978)	1112	112 (10.85271)	1032	1.089293	0.012
70+	70 (20.95808)	334	43 (18.61472)	231	1.060606	0.4822
Total	301 (11.85973)	2538	234 (9.458367)	2474	1.125988	0.0038

SDC: Spheroidal degeneration of cornea

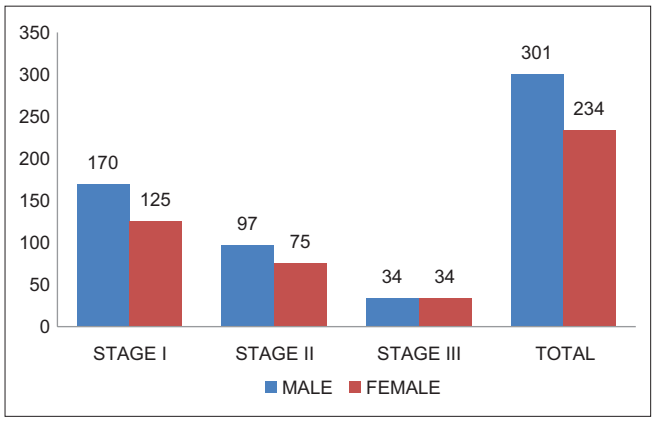

Figure 2: Distribution of spheroidal degeneration of cornea stages

Tables 3-5 list the "severity distribution" of SDC for Stage I, II, and III together, for Stage II and III together, and for Stage III alone. A total of 295 (55%) patients were found to have Stage I; 172 (32%) were found to have Stage II; and 68 (13%) were in Stage III, out of 535.

For women with SDC, the distribution of the stages is very similar in each of the four age groups. In Stage I, the percentage for the four age groups varied from 40% to 57.1%. In Stage II, the percentage varied from 27.9% to 40%, and for Stage III, the percentage varied from 14.9% to 23.2%.

In men, the percentage with Stage I was highest in the 40–50 years' age group (80.9%). In Stage I (male), the percentage of the other three age groups was almost similar, varying from 54.2% to 55.7%.

Older men have higher chances of developing Stage II and III than men of a younger age, the percentage range being 19.0% at 40–50 years' age group to 44.3%–45.8% for the 51–70 + age groups.

For women, the percentage developing Stages II and III does not increase with age significantly. The highest percentage is in the 40–50 years' age group (60%).

Age is thus an important factor in determining whether an affected man is likely to have progressed to Stage II and III, but this does not apply to women.

For Stage III alone, the percentage of women, in each group, was slightly higher except in those of 61–70 years of age than the corresponding percentage of men, but the difference was not significant.

Within both sexes, the percentage with Stage III does not vary significantly from one age group to another. The range for men was 4.8%–12.7% and for women was 10.7%–23.3%. Thus, neither age nor sex is a crucial factor in determining whether an individual with SDC is likely to have progressed to Stage III.

Association with other eye diseases

For each person in the sample, the presence or absence of arcus senilis, pterygium, cortical cataract, nuclear cataract, pseudoexfoliation, trachoma, and glaucoma was recorded. The association of each of these conditions with the prevalence of SDC was then examined.

Arcus senilis

The total number of SDC patients with arcus senilis was 283 and those in non-SDC group with arcus senilis numbered 2245. A person with arcus senilis is more likely to have SDC than a person without. However, the result is statistically insignificant ($P = 0.2187$).

Pterygium

A person with pterygium has three times greater risk of developing SDC than one without pterygium ($P < 0.0001$).

Cortical cataract

The prevalence of SDC was not significantly affected by the presence of cortical cataract (RR > 1).

Nuclear cataract

There was no association found between nuclear cataract and SDC (RR 0.8329).

Pseudocapsular exfoliation

A person with pseudocapsular exfoliation faces a 15 times higher risk of developing SDC and this difference was significant ($P < 0.0001$).

Glaucoma (primary open-angle glaucoma)

There was no association found between glaucoma and SDC.

Discussion

The prevalence of SDC in this study (10.674%) is perhaps higher than expected on clinical basis.

The increase in prevalence with age and the higher prevalence in men than women have been found in most samples, except in the Dahlak Island[4] and South-West Africa.[7] It has been explained on the basis of more and longer exposure to risk factors in elderly patients and in males who spend most of their time outdoors. In this population from

Table 3: Distribution of spheroidal degeneration of cornea for Stages I, II, and III

Age (years)	Stage I, *n* (%)	Stage II, *n* (%)	Stage III, *n* (%)	Total
Male				
40-50	17 (80.9524)	3 (14.28571)	1 (4.761905)	21
51-60	37 (54.4118)	24 (35.29412)	7 (10.29412)	68
61-70	77 (54.2254)	47 (33.09859)	18 (12.67606)	142
70+	39 (55.7143)	23 (32.85714)	8 (11.42857)	70
Total	170 (56.4784)	97 (32.22591)	34 (11.29568)	301
Female				
40-50	2 (40.00)	2 (40.00)	1 (20.00)	5
51-60	38 (51.3514)	25 (33.7838)	11 (14.86486)	74
61-70	64 (57.1429)	36 (32.1429)	12 (10.71429)	112
70+	21 (48.8372)	12 (27.907)	10 (23.25581)	43
Total	125 (53.4188)	75 (32.0513)	34 (14.52991)	234

Table 4: Distribution of spheroidal degeneration of cornea for Stages II and III

Age (years)	Males		Females	
	Stage II + III, *n* (%)	All stages (*n*)	Stage II + III, *n* (%)	All stages (*n*)
40-50	4 (19.047619)	21	3 (60)	5
51-60	31 (45.5882353)	68	36 (48.64865)	74
61-70	65 (45.7746479)	142	48 (42.85714)	112
70+	31 (44.2857143)	70	22 (51.16279)	43
Total	131 (43.5215947)	301	109 (46.5812)	234

South-West Rajasthan, women share outdoor occupations with men; women are doing most of the agricultural work: digging, planting, and cattle rearing, while men primarily look after the cattle. Both men and women do similar work in local cement factories. It is, therefore, reasonable to expect smaller differences between prevalence in male (11.86%) and female (9.458%) patients.

Analysis of the stages offers further explanation. The severity of SDC appears to be sex related because men have a slightly greater chance of developing Stage II and III, more so than women. So, some predominantly male factor or association may promote the progression of SDC once present. However, for Stage III alone neither age nor sex alter an affected person's chances of SDC. Having progressed to that stage, other factors must be important. These might be cultural, or involve the general health of the individual or a specific eye condition.

Vitamin and protein deficiencies and tuberculosis are common in this population.[8-10] Poor corneal nutrition may be a factor that increases the severity of degeneration, although it was not studied in this survey.[11]

There are few studies that have been conducted in the Indian subcontinent regarding the prevalence of corneal degeneration in different groups. Of the 10,293 participants in the Andhra Pradesh Eye Disease Study, only 0.064% (8) patients had SDC.[12] A study conducted by Singh and Bhullar

Table 5: Distribution of spheroidal degeneration of cornea for Stage III

Age (years)	Males		Females	
	Stage III, *n* (%)	Total	Stage III, *n* (%)	Total
40-50	1 (4.761905)	21	1 (20)	5
51-60	7 (10.29412)	68	11 (14.86486)	74
61-70	18 (12.67606)	142	12 (10.71429)	112
70+	8 (11.42857)	70	10 (23.25581)	43
Total	34 (11.29568)	301	34 (14.52991)	234

Table 6: Geographical features of South-West Rajasthan (districts Pali, Jalor, Sirohi, and Udaipur)[11]

Geographical features	Characteristics
Situation	24.20°-27.75° North latitude 72.16°-74.30° East longitude
Mean, annual bright sunshine	10-13 h/day
Temperature	Maximum: 32°C-45°C Minimum: 10°C-27°C
Rainfall, mean annual	40 cm/year
Humidity - average daily relative humidity near sunrise	25%-35%
Radiation, mean annual total (combined for direct and dispersed)	6.0-7.0 kWh/mm^2
Altitude	200-1000 m above sea level

in Punjab on the prevalence of SDC showed only 0.423%.[13] Both studies showed a very low prevalence rate. However, a study conducted by Viswamithra and Murthy showed higher prevalence (7.13%) similar to our results.[14]

The search for associations with specific eye diseases was not very rewarding. Pterygium, in which the etiology of UV irradiation is an important factor,[15,16] was twice as common in patients with SDC among the Dahlak Islanders[4] but not in the inhabitants of Arkansas,[7] and South African pondos.[2,4,6] In this study, pterygium was three times more common in patients with SDC.

Arcus senilis is a senile degenerative change which may be associated with either arterial disease or hypercholesterolemia. Association between arcus senilis and SDC was found in Dahlak Islanders[4] and South African pondos.[4,6] In this study, no statistically significant association was found between arcus senilis and SDC.

An association between SDC and glaucoma was found in Arkansas,[2] where 17% of patients with primary SDC had uncategorized glaucoma (57 out of 337) compared with 9.4% in the control group. It was concluded that glaucoma was an additional etiological factor. In this study, no relationship was found between glaucoma (POAG) and SDC.

There was a significant association between pseudocapsular exfoliation and SDC. A person with pseudocapsular exfoliation has 15 times higher risk of developing SDC. This agrees with Freedman's observation on the Namas of South-West Africa, where five patients with exfoliation were found in a group of 87 patients with SDC.[7]

Pseudocapsular exfoliation does share some common clinical features with SDC – increased prevalence with age and greater prevalence in males – but in the absence of a known etiology, neither a causal relationship nor common etiological factors appear likely, in spite of the finding of pseudocapsular-like material in extraocular sites, namely the conjunctiva.[17]

Prevalence of corneal degeneration, Stages II and III, in a country with a tropical climate is common. A geographical feature of etiological importance is the UV radiation level which is associated with extremes of temperature (high and low), low humidity, dust, and wind – the keratopathy of "tropical barren lands" and "climatic keratopathy."[2,4] South-West Rajasthan (Pali, Jalore, Sirohi, and Udaipur districts) has extreme climatic conditions. The area in which the survey was done was far from the coast and at an average height above sea level of 200 to 1000 m, with extremely arid climate [Table 6]. The area receives very low rainfall, and hot wind and dust storms occur in the summer. The landscape is covered with scrub jungles, and trees are scarce. The soil is sandy. The average maximum temperature in summer is 45°C and average minimum temperature is 10°C in winter. The area has low rainfall and about 325 days of good sunshine.[18]

One may speculate that chronic systemic infection (tuberculosis) and vitamin deficiency, both common in the population, might be factors in the production or progression of SDC and are worthy of future study. Public education on the care of eyes needs to be conducted and field workers should be advised to wear goggles.

Conclusion

Extreme temperature, low humidity, dust, high wind, and microtrauma caused by sand particles may be the probable etiologies for higher prevalence of SDC in the tribes of western Rajasthan. Pterygium and pseudocapsular exfoliation seem to be the associated ocular comorbidities.

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Conflicts of interest

There are no conflicts of interest.

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