Original Research Article

Anterior segment findings and systemic disease associations in scleritis

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Abstract

Objectives: To evaluate Anterior segment findings and systemic disease associations in patients with scleritis and to assess any ocular or systemic prognostic significance of it. **Materials and Methods:** Patients with scleritis alone and patients with scleritis-associated peripheral keratopathy; attending our hospital during period of January 2011 to January 2015 were examined. Review of patients with scleritis and the different patterns of peripheral keratopathy: (peripheral corneal thinning, stromal keratitis, and peripheral ulcerative keratitis (PUK)), review of ocular and systemic outcomes were done and comparisons between patients with scleritis with and without peripheral keratopathy was done. **Results:** Patients with scleritis associated with Keratopathy had significantly more necrotizing scleritis (P<.05), decrease in vision (P<.05), anterior uveitis (P<.05), and potentially lethal specific-disease association (P<.05) than did patients with scleritis alone. Patients with Peripheral keratopathy had the worst ocular and systemic outcomes. Of the 24 patients with Peripheral keratopathy 58% had necrotizing scleritis (P<0.05), virtually all had a potentially lethal systemic disease (P<0.05). **Conclusion:** The detection of peripheral keratopathy, and especially in a patient with scleritis indicates a poor ocular and systemic prognosis.

Key Words: Anterior segment, scleritis.

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INTRODUCTION

Keratopathy associated with scleritis usually occurs directly adjacent to or in the same quadrant as the active scleritis. It may progress circumferentially, centrally, or posteriorly (in which case corneal perforation may occur). Early diagnosis and treatment may improve the ocular prognosis before vision is affected or perforation is imminent. In a study of enucleated eyes with scleritis associated with rheumatoid arthritis, Sevel[1] found that 36% had peripheral keratopathy, and in different clinical studies, Watson *et al*[2,3] found that 29% to 37% of patients with scleritis had peripheral keratitis. Peripheral keratopathy associated with scleritis may be relatively benign or serious, and clinical differentiation of the types of keratopathy may be important if they have different ocular and systemic prognostic significance. The clinical types of peripheral keratopathy associated with scleritis include peripheral corneal thinning (intact epithelium and no inflammatory cells in the stroma), stromal keratitis (intact epithelium but with inflammatory cells in the stroma and without stromal ulceration), and peripheral ulcerative keratitis (PUK) (epithelial defect, inflammatory cells in the stroma, and stromal ulceration). Peripheral keratitis with scleritis has varying degree of vasoocclusive changes in episcleral and conjunctival vasculature which may lead to impending corneal perforation due to resorption of stroma[3,4,5].Vaso occlusion may be due to inflammatory microangiopathy as an extension of systemic vasculitis process or local immune response[8]. This study evaluated the ocular characteristics and systemic disease associations of patients with all three patterns of scleritis-associated peripheral keratopathy.

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MATERIALS AND METHODS

We reviewed 248 consecutive patients with scleritis seen in KIMS OPD during four year of duration. The patients were divided in two groups- 1) Those having only scleritis 2) Those having keratopathy. Patients with scleritis-associated with peripheral keratopathy (SAPK) were further sub divided depending on the type of keratopathy as five types.

Peripheral corneal thinning (PCT): The periphery of the cornea (within 2-3 mm of the limbal margin) becomes grayish and about one-third thinner in one or more areas. The epithelium remains intact throughout the thinning process.

- **Stromal keratitis:** Isolated or multiple white or gray nummular opacities composed of leukocytes within the corneal stroma. The epithelium is intact.
- **Peripheral ulcerative keratitis (PUK):** Peripheral corneal ulcer with epithelial defects, stromal infiltration, and stromal degradation. The ulcer has a well-defined border on its limbal and corneal edges.
- **Sclerosing Keratitis**: Keratitis adjascent to inflammaed sclera characterized by corneal opacification and varying degree of vascularization. No epithelial defect.
- Others: Limbal guttering and preexisting opacities

MATERIAL AND METHODS

All preliminary Ophthalmological examination along with slit lamp examination was carried out. Patients data including age, sex, bilaterality, type of scleritis, type of peripheral keratopathy (SAPK,) history of previous ocular surgery associated anterior uveitis, Systemic disease association, were analyzed. Scleritis was characterized as anterior or posterior, according to the classification of Watson and Hayreh[2]. Anterior scleritis included the diffuse, nodular, necrotizing, and scleromalacia perforans types. Ocular manifestations, including loss of best-corrected visual acuity, anterior uveitis, and impending corneal perforation, were analyzed. Decrease in visual acuity was defined as loss of vision of two or more Snellen lines at the end of the follow-up period or visual acuity of 20/80 or worse at presentation related to the inflammatory disease process. Anterior uveitis was diagnosed based on the detection of inflammatory cells in the anterior chamber with or without synechiae or keratic precipitates. Impending corneal perforation was defined as progressive destruction of the peripheral cornea, leaving only some layers of deep stroma and/or Descemet membrane**.** Association of various spondyloartropathies (Rheumatoid arthritis, Polyarthritis nodosa, Spondylising arthritis, Wagner's) were noted. All patients were examined by rheumatologist and necessary specific investigations were advised which include compatible history, review of system, laboratory and biopsy data. Data were analyzed with a customized database software program, Comparisons were made between patients with SAPK and scleritis alone. Comparisons were also made among scleritis patients with different patterns of peripheral keratopathy and sysyemic associations. Statistical analysis was performed with the χ^2 test.

RESULTS

Total 248 patients of scleritis were examined out of which 168(67.74%) had only scleritis while 80(32.26%) had associated keratitis (SAPK).

Table 1:

Scleritis	No.	%
Scleritis alone	168	67.74
Keratitis	80	32.26
Total	**248**	

Out of 248 patients, male to female ratio is 0.95, which is similarly distributed among patients having scleritis alone and SAPK

Table 2:

Sex	Scleritis		Keratitis		Total	
Male	82	48.8%	39	48.7%	121	48.8%
Female	86	51.19%	41	51.3%	127	51.2%
Total	**168**		**80**		**248**	

42 (16.94%) patients had bilateral scleritis and 206 (83.06%) had it unilateral. Out of 42bilateral 26 (61.9%) had scleritis alone and 16 (38.1%) had associated keratopathy.

Table 3:

Laterality	Scleritis		Keratitis		Total	
Bilateral	26	15.48%	16	20%	42	16.94%
Unilateral	142	84.52%	64	80%	206	83.06%
Total	**168**	**100%**	**80**	**100%**	**248**	**100%**

60% patients having keratopathy had Anterior uveitis ($P < 0.05\%$).

Table 4:

Anterior Uveitis	Only scleritis	Scleritis and Kpathy
Present	34	48
Percentage	20.24%	60%

Scleritis patients (248) were sub divided into Diffuse (56,22.58%), Nodular (90,36.29%), Necrotizing (78, 33.06%), Scleromalacia (16, 6.45%), Ant. and Post. Scleritis (8, 3.23%).

Table 5:

Scleritis	No.	%
Diffuse	56	22.58
Nodular	90	36.29
Necrotising	78	33.06
Scleromalacia	16	6.45
Ant. and post scleritis	8	3.23
Total	**248**	

Patients having different types of posterior scleritis had keratitis are 17.8% of Diffuse, 31.1% of Nodular and 48.7% of Necrotizing had keratitis, 25% of having Scleromalacia.none of the patient had keratitis having anterior and posterior scleritis.

Table 6:

Type of Scleritis	Total	Keratitis	%
Diffuse	56	10	17.8
Nodular	90	28	31.1
Necrotising	78	38	48.7
Scleromalacia	16	4	25
Ant. and post scleritis	8	0	0
Total	**248**	**80**	

Most common keratopathy noted was Stromal (55%), then PCT (17.5%), PUK (12.5%), Sclerosing keratitis (10%), and others (5%) in descending order.

Table 7:

Type of Keratitis	No.	%
PUK	10	12.5
Stromal	44	55
PCT	14	17.5
Sclerosing Keratitis	8	10
Others	4	5
Total	**80**	

Patients having Necrotizing scleritis had higher prevalence of keratopathy (48.72%), and then Nodular (31.11%), Scleromalacia (25%), and diffuse (17.86%) in descending order. ($P < 0.05$)

Table 8:

Type of Scleritis	Total	Scl,Kpathy	Total
Diffuse	46	10, 17.86%	56
Nodular	62	28, 31.11%	90
Necrotising	40	38, 48.72%	78
Scleromalacia	12	4, 25%	16
Ant. and post scleritis	8	0	8
Total	**168**	**80, 32.26%**	**248**

Type of keratopathy most common was stromal and peripheral corneal thinning in Necrotizing and Nodular scleritis. ($P< 0.05$).

Table 9:

Type of scleritis	PUK	Stromal	PCT	Sclerosing Keratitis	Others	Total
Diffuse	2, 20%	8, 18.2%	0	0	0	10
Nodular	2, 20%	14, 31.8%	6,42.8%	6, 75%	0	28
Necrotising	6, 60%	18, 40.9%	8, 57.14%	2,25%	4, 100%	38
Scleromalacia	0	4, 9.1%	0	0	0	4
Ant. and post scleritis	0	0	0	0	0	0
Total	**N=10,**	**N=44**	**N=14**	**N=8**	**N=4**	**80**

Rheumatoid Arthritis (6.41%) was the most common associated systemic disease, out of which 37.5% had keratopathy. Next common was Wegener Granulomatosis (4.84%) and 67.5% had keratopathy ($P < 0.05$).

Table 10:

	Scleritis	Scleritis and,kpathy	Total
Rheumatoid arthritis	18, 50%	10, 38.5%	28,11.3%
Spondylo arthritis	2,	0	2,0.8%
Relapsing polychondritis	2,	0	2,0.8%
Wegener granulomatosis	4,11.1%	8,30.76%	12,4.8%
Polyarteritis nodosa	0	4,15.4%	4,1.61%
Infectious	8,22.2%	2,	10,4%
Others	2	2,	4,1.6%
Total	**N=36**	**N=26**	**N=248**

Table 11:

	Average ESR	p value
Scleritis, systemic association	61.17(±35.27)	
Scleritis without systemic association	47.56(±18.16)	0.05
Scl, kert, syst	75.38(±47.37)	
Scl, kert, without systemic	49.56(±21)	0.01

DISCUSSION

The results of these study reveal severity of scleritis associated with keratopathy. Scleritis associated with peripheral keratopathy can be regarded as bad prognostic sign because these patients more often had necrotizing scleritis, decrease in vision, anterior uveitis, and impending corneal perforation. Our proportion of scleritis patients with peripheral keratopathy (32.26%) is comparable to study performed in 1976 by Watson and Hayreh[2] at Moorfields Eye Hospital, London, England (88 of 301 eyes, 29%). The detection of peripheral keratopathy in a patient with scleritis is also a warning sign of increased likelihood of an associated disease[1]. In this study, scleritis patients with peripheral keratopathy more often had an associated disease than patients with scleritis alone (68.82% vs31.18%). Rheumatoid arthritis, Wegener granulomatosis, and infectious diseases were the most common associated diseases. Several distinctive patterns of peripheral keratopathy are found in patients with scleritis[6,7]: PCT, stromal keratitis, and PUK. In our experience, all three patterns were seen to cause decrease in vision and associated with anterior uveitis.60% of PUK had necrotizing scleritis, 57.14% of PCT had necrotising sceritis, both had grave visual prognosis.

CONCLUSION

Patients having Necrotizing and Nodular scleritis are more prone to have Stromal keratitis and PCT. Necrotising scleritis had more association with PUK and PCT which has grave prognosis. Close follow up is always required for all patients having Stromal keratitis and PCT for any signs of scleritis. In all patients with peripheral keratopathy and scleritis search must be made for an underlying disease by doing necessary investigations. Scleritis patients with peripheral keratopathy are likely to have an associated systemic disease, the most common being rheumatoid arthritis[1], Wegener granulomatosis, polyartitis nodosa and infectious diseases.

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