ORIGINAL PAPER DOI: 10.5603/OJ.2022.0013

Clinical profile of patients with the first presentation as anterior nodular scleritis — a case series

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ABSTRACT

BACKGROUND: The aims of this study was to report the clinical profile of five patients with features of anterior nodular scleritis as initial presentation in residents of Western Maharashtra.

MATERIAL AND METHODS: A retrospective review of medical records was done from February 2019 to December 2019. Five new anterior nodular scleritis cases were reported at the tertiary-eye care centre within two months. The slit-lamp examination, anterior-segment photographs, and laboratory data were collected and analysed.

RESULTS: A majority of cases (4/5) were presented in females. Three patients had a unilateral presentation, while 2 — had bilateral involvement. The most common presenting symptom was ocular pain and redness. Corneal involvement was seen in 1 case. Two female patients were diagnosed with rheumatoid arthritis, one female had multinodular goitre, and the other two were idiopathic. Three patients responded very well to oral non-steroidal anti-inflammatory drugs (NSAIDs), while the rest two were managed with steroids. No patient had to be started on immunosuppressives. All the cases showed complete resolution with the treatment provided. All these patients presented at our centre in the autumn season. All these patients presented at our center in the autumn season only and no new case of anterior scleritis reported before or after this season.

CONCLUSION: Management of scleritis may need a multidisciplinary approach as it may be the only presentation of an underlying systemic disease. It is to be considered in the differentials of red eye that presents with a substantial amount of ocular pain or with the known systemic disorder that leads to scleritis development. Also, there could be a seasonal association of anterior-nodular scleritis like episcleritis.

KEY WORDS: anterior scleritis; rheumatoid arthritis; seasonal variation; autumn season; nodular

Ophthalmol J 2022; Vol. 7, 80–85

INTRODUCTION

Scleral inflammatory diseases consist of episcleritis and scleritis. Scleritis is an ocular inflammatory disorder with a broad spectrum of disease and presentation. It causes disabling ocular pain and visual disturbance [1]. Approximately half the patients affected have no identifiable cause, but 30–40% have an associated systemic autoimmune condi-

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tion [1]. Scleritis may be the initial presentation of many systemic conditions, and thus, all patients with scleritis require a thorough systemic evaluation [1]. The occurrence of scleritis in known cases of rheumatoid arthritis has been reported from 0.3% to 0.67% [2–4]. Anterior scleritis is sub-classified as diffuse, nodular, and necrotizing. The majority of the scleritis cases develop in patients in the fourth-to-sixth decade of life [5]. Though the occurrence of episcleritis has been shown more frequently in the autumn season than in summer and winter, scleritis also might have a seasonal variation [6]. The highest incidence of scleritis in males occurred in the fourth decade, while females had two peaks: in the third and sixth decade of their lives [6]. Studies report that in women, necrotizing scleritis ranges from 66% to 83% [7–11]. Bilateral presentation of scleritis is more commonly seen in patients with the underlying systemic disease [12–14]. The ocular inflammation is often severe and sometimes requires immunosuppressives. Next to the diffuse type, nodular scleritis is the most common subtype of anterior scleritis. Nodular scleritis creates a diagnostic challenge for ophthalmologists because of its various differentials.

The available literature has explained the clinical profile of patients with scleritis, but there is a lack of literature on isolated cases of anterior nodular scleritis. Here we report 5 cases with the first presentation as anterior nodular scleritis. Our series describe the clinical profile, approach, and management of new cases of anterior nodular scleritis in the Western population.

MATERIAL AND METHODS

It was a retrospective case series. All the cases with anterior nodular scleritis reported at the tertiary eye care centre from February to December 2019 were enrolled in the study. The study was approved by the local Institutional Ethical Committee and was carried out according to the Ethical principles of the Declaration of Helsinki. All the cases of anterior nodular scleritis were included in the study as per classification by Watson and Hayreh. These were diagnosed based on clinical signs such as nodular swelling over the sclera and congestion of deep episcleral vessels with or without severe pain. Blanching test with phenylephrine confirmed congestion of deep episcleral vessels. Detailed anterior and fundus examination was noted. Patients underwent detailed laboratory evaluation, including complete blood count, rheumatoid factor, and antinuclear cytoplasmic antibody. A multidisciplinary approach was implemented, and rheumatologist opinions were taken in required cases. All the patients were managed with topical and oral non-steroidal anti-inflammatory agents, and one case was on topical corticosteroid as well. Patients who did not respond to this treatment were given oral corticosteroids after the rheumatologist's opinion. Oral corticosteroid was started at 0.5–1 mg per kilogram of body weight per day and was tapered slowly based on the clinical recovery. The resolution was well-defined as no ocular symptom, along with the resolution of clinical episcleral congestion, scleral nodule, and edema. Exclusion criteria included other subtypes of scleritis, inadequate information and data available, trauma, and past extra-ocular surgery.

Case 1

A 45-years-old female presented with a complaint of excruciating pain in both eyes of 7 days duration. She reported a history of occasional joint pain. On examination, her unaided distant visual acuity was 20/20 in both eyes. Slit-lamp examination showed localised conjunctival congestion with prominent and tortuous episcleral and scleral vessels mostly involving the temporal portion in both eyes. Cornea, anterior chamber, lens, and fundus examination were all within normal limits. Intraocular pressure was normal in both eyes. 10% phenylephrine caused blanching of superficial vessels but not the deep vessels, confirming scleritis. USG B-scan showed the normal thickness of the sclera at the posterior pole, ruling out posterior scleritis then and there. The patient was diagnosed with anterior scleritis and started on topical loteprednol (drops Lotepred 0.5%) 6-hourly, topical flurbiprofen (drops 0.03%) 8-hourly, and indomethacin (tablet Indomethacin SR 75 mg) 24-hourly. The patient was advised to perform blood investigations. On follow-up after two weeks, the patient was clinically and symptomatically better. Investigation reports showed blood count and erythrocyte sedimentation rate (ESR) within normal limits. On further follow-up, ocular examination showed scleral thinning in the temporal area in both eyes with inflammation. Investigations revealed elevated levels of C-reactive protein (CRP), and rheumatoid arthritis (RA) factor was 20.16 IU/mL (normal < 14 IU/mL). The diagnosis was then changed to necrotising anterior scleritis secondary to suspected RA. The pa-

tient was immediately referred to a rheumatologist, and the oral steroids were started in tapering doses. After one month of treatment, she was symptomatically much better. Ocular pain and joint pain had resolved, and the eye looked quiet.

Case 2

A 54-years-old female, with the known history of multinodular goitre, presented with complaint of pain and redness in the right eye lasting for six days. On examination, her unaided visual acuity was 20/40 in both eyes, improving to 20/20 with glasses correction. Slit-lamp examination revealed nodular swelling with prominent and tortuous episcleral and scleral vessels temporally to the cornea in the right eye. Other anterior segment examination and fundus examination were normal. Intraocular pressure was 14 mm Hg in both eyes. 10% phenylephrine did not cause blanching of prominent deep vessels over the nodule, confirming scleritis. USG B-scan ruled out posterior scleritis. The patient was diagnosed with nodular anterior scleritis **[in the?]** right eye and started on oral ibuprofen 400 mg 8-hourly and topical ketorolac 0.5% 8-hourly. Blood counts, ESR, CRP, and veneral diseases research laboratory (VDRL) tests were within normal limits on investigations. RA factor was also within the normal limit. The patient improved symptomatically.

Case 3

A 28-years-old female presented with a complaint of severe pain and redness in the left eye lasting for three days. Unaided visual acuity was 20/20 in both eyes. On slit-lamp examination, the cornea was clear. There was a nodular swelling temporally to the cornea associated with prominent tortuous scleral vessels. Intraocular pressures were 12 and 14 mm Hg in the right and left eyes, respectively. 10% phenylephrine caused blanching of engorged superficial vessels, but deep vessels were still prominent. USG B-scan ruled out posterior scleritis by showing normal sclera thickness at the posterior pole. The patient was diagnosed with nodular anterior scleritis of the left eye and started on oral indomethacin (Indomethacin SR 75 mg) 12-hourly, and topical ketorolac 0.5% 8-hourly. On investigation of blood counts, ESR was within normal limit. CRP and RA factor tests were negative. Chest X-ray was normal. VDRL test was negative. Follow-up after one week showed a significant reduction in pain and redness. The patient was advised to continue the treatment for three more weeks and continue with topical medications thereafter.

Case 4

A 33-years-old male presented with pain in the right eye lasting for five days. He reported also a headache in the right temporal region. On examination, his uncorrected visual acuity was 20/20 in both eyes. Slit-lamp examination showed nodular elevation temporally to the cornea with prominent episcleral vessels above the nodular swelling and prominent bluish-red scleral vessel beneath those episcleral vessels. The rest of the anterior segment, fundus examination, and intraocular pressure were within normal limits. 10% phenylephrine did not affect the engorged deep vessels. However, the superficial episcleral vessels were blanched. USG B-scan was performed to rule out posterior scleritis. The patient was diagnosed with nodular anterior scleritis with episcleritis of the right eye. The patient was started on oral ibuprofen 400 mg 8-hourly, oral pantoprazole 40 mg 24-hourly, and topical ketorolac 0.5% 8-hourly. Investigations showed blood counts within normal limits. ESR was 15 mm/hr, CRP test was negative, RA factor was within normal limit, and anti-nuclear antibody (ANA) test was negative. Chest X-ray was normal. On follow-up, after two weeks, the patient improved symptomatically, and examination showed a decrease in episcleritis and scleritis (Fig. 1). The patient was advised to continue the same treatment for two more weeks. He then continued treatment with topical ketorolac for two more weeks.

Case 5

A 50-years-old female presented at our centre with pain and redness in the right eye lasting for two weeks. The patient had visited another facility earlier and was prescribed topical steroids for the last ten days. The redness, however, was unresolved, and the pain in her right eye was still lasting. On examination in our centre, the patient's uncorrected visual acuity was 20/60, improving to 20/40 with a pinhole in both eyes. On slit-lamp examination of the right eye, there was conjunctival congestion temporally and grade II nasal pterygium. There was nodular swelling of the sclera and prominent episcleral and scleral vessels temporally. Cornea showed thinning of 2.5 x 2.5 mm, located paracentrally, not involving visual axis and temporally shown in Figure 2. The anterior chamber was within the normal limit. The lens showed nuclear sclerosis of grade I, and fundus evaluation was

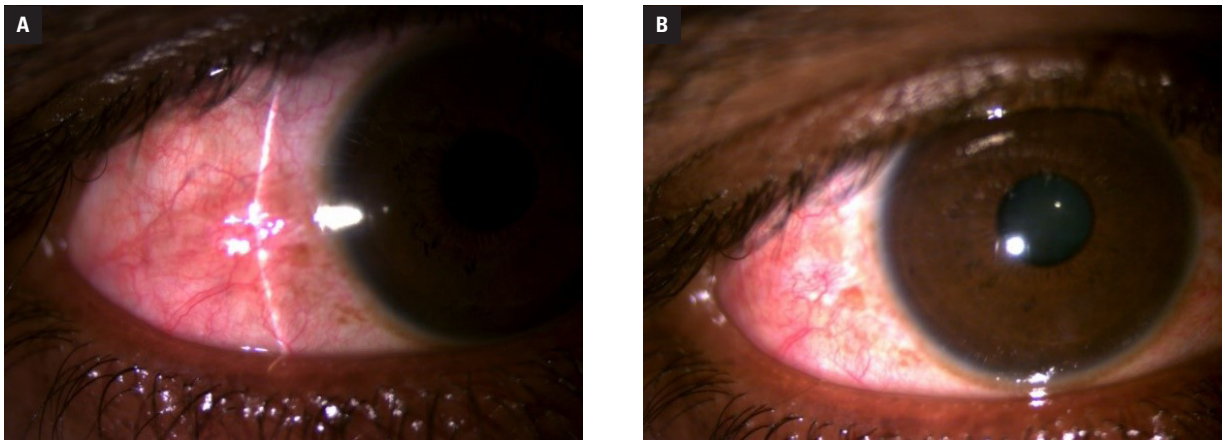

FIGURE 1. A. Nodular anterior scleritis showing nodular elevation of sclera associated with prominent episcleral and scleral vessels; **B.** After 7 days of treatment, decrease in amount of nodular swelling and vascular congestion as in Case 4

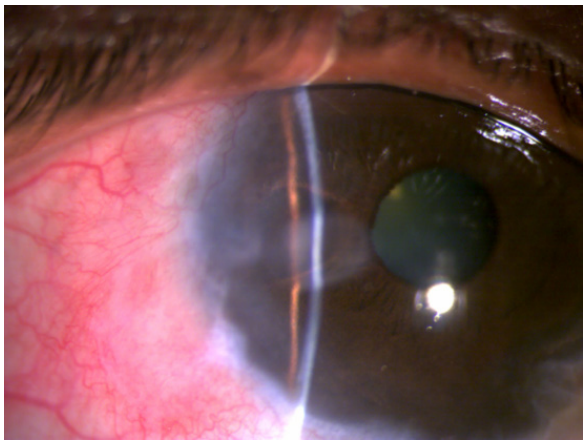

FIGURE 2. Nodular anterior scleritis with dellen right eye. Picture showing dellen on the cornea, nodular swelling with prominent and tortuous diffuse vessels on the sclera associated with conjunctival congestion as seen in Case 5

within normal limits. Examination of the right eye showed grade II nasal pterygium and grade I nuclear sclerosis. IOPs were normal in both eyes. 10% phenylephrine did not blanch the engorged deep vessels over the nodule. USG B-scan ruled out posterior scleritis. In view of dellen and scleritis, the treatment was revised, and the patient was started on oral prednisolone 50 mg 24-hourly, indomethacin (tablets) 12-hourly, and topical ketorolac 0.5% 6-hourly. Blood counts were within the normal limit. ESR was 28 mm/hr. RA factor and CRP tests were positive. The patient was then referred to a rheumatologist for an opinion. Oral steroids were continued in tapering doses along with analgesics. The patient responded well to the treatment with complete resolution of anterior scleritis.

RESULTS

The demographic factors and clinical profile are summarized in Table 1.

DISCUSSION

Diffuse anterior scleritis is the most common, and scleromalacia perforans is the least common type of scleritis [6, 15, 16]. Non-necrotizing scleritis usually has a gradual onset of ocular pain which can be localized or radiating. Necrotizing scleritis is far more devastating than the non-necrotizing type due to the severity of its likely sequelae together with vision loss [6–9]. Necrotizing anterior scleritis with inflammation (as in Case 1) usually manifests with gradual onset of photophobia. Severe pain from all types of scleritis can cause disturbance in regular sleep habits and lead to weight loss [6, 9]. A grey or blue-grey appearance of the sclera may push the patient to seek treatment [17]. Scleritis is associated with an underlying systemic condition in up to 57% of the cases (as in Cases 1 and 5), which is usually autoimmune [18]. A patient may also report a history of systemic symptoms like joint pain (as in Case 1). In our case, it was strongly associated with RA. Scleromalacia perforans is almost always bilateral and seen in patients with long-standing RA. So, we counseled our first and fifth patients with RA about the ocular complications they could face during their lifetime. Nodular scleritis can be idiopathic [19] (as in Cases 2–4).

In our case series of 5 patients with scleritis, 4 were females, and one was male. Two females were in the fifth decade, one in the fourth decade, and one in her late twenties. The male patient was

Table 1. The demographic factors and clinical profile of new cases who presented with anterior scleritis as their first presentation

Case	Age/ Sex	First symptom	BCVA both eyes	Comorbidities	Topical [eye drops]	Systemic [tablets]	Response
1	45/F	Pain lasting 7 days, both eyes	20/20	RA	Loteprednol 0.5% Flurbiprofen 0.03%	Indomethacin 75 mg Prednisolone 50 mg, with tapering dose	Complete resolution
2	54/F	Pain and redness lasting 6 days, right eye	20/20	Multinodular goitre	Ketorolac 0.5%	Ibuprofen 400 mg	Complete resolution
3	28/F	Pain and redness lasting 3 days, left eye	20/20	None	Ketorolac 0.5%	Indomethacin 75 mg	Complete resolution
4	33/M	Pain and redness lasting 5 days, right eye	20/20	None	Ketorolac 0.5%	Ibuprofen 400 mg	Complete resolution
5	50/F	Pain and redness lasting 14 days, right eye	20/20	RA	Preservative-free lubricant Ketorolac 0.5%	Indomethacin 75 mg Prednisolone 50 mg with tapering dose	Complete resolution

F — female; M — male; BCVA — best-corrected visual acuity; RA — rheumatoid arthritis

in his mid-thirties. The age at presentation in both genders was almost similar to that reported in Watson's and Hayreh's study. Unlike scleritis, episcleritis occurs more commonly in the spring and autumn season. In our centre, all the new cases of anterior scleritis presented during the autumn season only, and there were no new cases of scleritis apart from this period. This seasonal variation might be there and apt for scleritis too. This might help keep anterior scleritis as differential in patients that present with red eye and ocular pain during the autumn season.

There is an associated episcleritis in all types of scleritis — except scleromalacia perforans. In our cases, most patients had scleritis associated with episcleritis. A conclusive diagnostic approach is to use 10% phenylephrine, which blanches conjunctival and episcleral vessels and leaves the scleral blood vessels engorged. Corneal involvement or sclerokeratitis establishes as infiltrative stromal keratitis, non-inflammatory corneal thinning, limbal guttering or peripheral ulcerative keratitis (PUK) [17, 20]. As in Case 5, there was corneal thinning seen adjacent to scleritis. The association between nodular scleritis and systemic rheumatic disorders ranges from 0% to as high as 45% [21]. In a retrospective analysis, Raiji et al. observed that patients with systemic diseases were more likely to develop diffuse or necrotizing scleritis, whereas those with isolated scleritis had nodular scleritis [22]. In this case series of 5 patients, RA was observed in 2 cases, 1 case was associated with multinodular goitre, and the rest were found to have idiopathic causes.

Oral non-steroidal anti-inflammatory drugs (NSAIDs) are the first line of scleritis treatment, and they are efficacious in treating non-necrotizing scleritis [21–23]. It has been suggested that indomethacin and oxyphenbutazone are the most effective. This approach usually controls non-necrotizing anterior scleritis (as seen in Cases 2–4). Oral steroids are usually considered if there is associated anterior uveitis or if NSAIDs are ineffective (as seen in Cases 1 and 5). In our study the patients who were comorbid with RA had to be managed on oral steroids as their symptoms were more severe. All the patients recovered well without using immunosuppressive drugs, and there was no recurrence. Our results suggest that new cases of anterior nodular scleritis may be very well treated with oral NSAIDS or sometimes steroids. Case 1 patient did not improve with oral NSAIDs and was later started on oral steroid and referred to a rheumatologist after her investigations were suggestive of RA.

Though our study is limited by its retrospective design and a small number of cases, it has many strengths. There is a scarcity of literature describing the association between seasonal variation and nodular scleritis. Also, not many studies have described nodular scleritis alone as the majority of case series have shown it as a subset of scleritis only. To the best of our knowledge, this is the one series that is utterly based on anterior nodular scleritis alone presented during the autumn season. Studies with a larger population are warranted.

CONCLUSION

Scleritis can be the only presentation of severe systemic disease. While scleritis is rare, it is undoubtedly more common than reported due to underdiagnosis. Scleritis should be considered one of the differential diagnoses of red eye that presents with a substantial amount of ocular pain or any known systemic disorder capable of producing scleritis. Also, there could be some seasonal association with the onset of this disease, like episcleritis. So, further studies can be carried out to support the relationship between the onset of this disease with seasonal factors.

Conflicts of interest

None declared.

Financial support

None declared.

Acknowledgement

None declared.

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