Original Article

Clinical profile of patients with posterior scleritis: A report from Eastern India

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Purpose: This study aimed to report the clinical profile of patients with posterior scleritis at a tertiary eye center in Eastern India. **Methods:** This was a single-center retrospective case series of patients who were diagnosed as posterior scleritis between January 2010 and December 2014, with a follow-up period of at least 6 months. **Results:** The study included 18 patients of posterior scleritis with a mean age of 41.2 ± 10.6 years (range: 26–63 years). With female preponderance (55.6%), majority of the posterior scleritis cases were unilateral (88.9%). Sixteen patients reported with diminution of vision, eleven patients (61.1%) had ocular pain on presentation, and five patients complained of headache. Concurrent anterior scleritis was found in three eyes (15%) with posterior scleritis. Choroidal folds and subretinal fluid at the posterior pole were the most common fundus findings and were seen in seven eyes (35%) each. No systemic association was detected in any patient even after extensive laboratory workup and multidisciplinary consultation. All patients received oral steroid, and 11 (61.1%) of them required intravenous pulse steroid therapy. Immunosuppressive was used in 6 (33.3%) patients, and oral azathioprine was the most common immunosuppressive used in the study. Recurrence was noted in eight eyes (40%). The mean best-corrected visual acuity improved to logarithm of the minimal angle of resolution (logMAR) 0.06 ± 0.051 at the final follow-up from 0.47 ± 0.45 logMAR at presentation (P = 0.00608). **Conclusion:** Posterior scleritis is relatively rare but can occur without systemic involvement. Aggressive immunomodulatory therapy is required to treat vision-threatening condition.

Key words: Immunosuppressive, posterior scleritis, scleritis

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Posterior scleritis refers to the inflammation of the sclera beyond equator.[1,2] Posterior scleritis has been reported to account for 2%–12% of all the cases of scleritis in literature.[2] Unlike other subtypes of scleritis, posterior scleritis has relatively lower systemic association.[2] Posterior scleritis is known for vision-threatening presentations and often needs aggressive therapy to prevent vision loss.[3]

Due to its plethora of presentations and rare occurrence, diagnosis of posterior scleritis remains largely eluded. Many of the times diagnosis of posterior scleritis is not considered or posterior scleritis is misdiagnosed as some other clinical entity.[2] This may be a reason for relatively sparse literature on posterior scleritis from India. The current study was conducted in a tertiary eye center from Eastern India to understand the clinical profile of the patients with posterior scleritis.

Methods

This was a hospital-based retrospective case series, which analyzed all the consecutive patients with posterior scleritis presenting to a tertiary eye care center between January 2010 and December 2014. The study was approved by the Institutional Review Board of our hospital, and all the research was performed in adherence to the tenets of the Declaration of Helsinki.

All patients receiving a clinical diagnosis of posterior scleritis were included in the study. Posterior scleritis was diagnosed based on the clinical and ultrasonographic findings. Clinical features such as ocular pain, worsening on eye movement, and decrease of vision with hyperopic shift in refractive error and clinical findings in the posterior segment on fundus examination supported by posterior scleral thickening and presence of sub-Tenon's fluid on ultrasonography B-scan helped us to reach a diagnosis of posterior scleritis. Patients who had been previously diagnosed or treated for posterior scleritis were excluded from the study.

A detailed ocular and systemic history was obtained for each patient, and history of Koch's contact was stressed and asked for in each patient. After taking a detailed medical history, each patient underwent a complete eye examination including assessment of best-corrected visual acuity (BCVA), slit-lamp biomicroscopy, and fundus examination with +90/+78D and indirect ophthalmoscopy. Laboratory investigation performed in these patients included a complete blood count, C-reactive protein, serum angiotensin-converting enzyme (ACE), tuberculin skin test, chest X-ray, rheumatoid factor (RF), antinuclear antibody, human leukocyte antigen (HLA) B-27, cytoplasmic and perinuclear antinuclear cytoplasmic antibody, venereal disease research laboratory test, and

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Treponema pallidum hemagglutination assay. Investigation such as high-resolution computed tomography (HRCT) chest was used in specific cases to rule out the presence of systemic granulomatous disease. Data retrieved from the patient's clinical records, including sociodemographic factors, medical history, clinical, laboratory and ultrasound findings, and treatment outcome, were entered into a computerized database. Furthermore, details of topical drugs as well as systemic corticosteroids and immunosuppressive agents were recorded. BCVA results were converted to logarithm of the minimal angle of resolution (logMAR) for statistical analysis and are presented as logMAR and Snellen equivalent. Ocular hypertension was defined as intraocular pressure >20 mmHg. Improvement of visual acuity was defined as an increase in BCVA by two lines or more in Snellen chart. Statistical analysis was performed using IBM SPSS Statistics, version 20.0 (International Business Machine Corp., Armonk, NY, USA), and $P < 0.05$ was considered statistically significant.

Results

Eighteen cases of posterior scleritis were seen at the tertiary eye center at Eastern India between January 2010 and December 2014. The main characteristics of these patients are presented in Table 1. Eight patients (44.4%) were male and 10 were female (55.6%). Mean age at presentation was 41.2 ± 10.6 years (range: 26–63 years). Diminution of vision was the most common complaints reported by all of our patients. Ocular pain was the presenting complaints in 11 patients (61.1%), and five patients (27.8%) also complained of headache at the time of presentation. One patient with unilateral posterior scleritis presented with eyelid edema. Nine eyes (45%) showed anterior chamber reaction of varying degree. Three patients (15%) with unilateral posterior scleritis had anterior scleritis at the time of presentation. Choroidal folds and serous retinal detachment were the most common clinical signs on fundus examination and were seen in seven eyes each (35%). Five eyes (25%) had disc edema, and one of them had macular star such as arrangement of hard exudates in fovea. These patients were evaluated extensively, and any possibility of neurological involvement was excluded after detailed examination. One patient presented with unilateral subretinal mass and was found to have giant nodular posterior scleritis on subsequent evaluation.

All of our patients had idiopathic posterior scleritis. Despite extensive investigation and multidisciplinary approach which included consultation from a rheumatologist and in-house physician, the cause of scleral inflammation in patients of the current study remained idiopathic. One patient was tested positive for RF, but no evidence of systemic disease was observed. Raised serum ACE and positive interferon-gamma release assay were found in one patient each, but HRCT of the chest was unremarkable in them. Ultrasound B-sonography showed thickening of the outer coat of the eyeball in 16 eyes (80%), and 6 eyes showed the presence of T-sign on ultrasound [Fig. 1].

Topical steroid with cycloplegic was used to control anterior uveitis, and all the patients received oral steroid (1 mg/kg/day in tapering schedule). Intravenous pulse corticosteroid (intravenous methylprednisolone) was administered in 11 patients (61.1%) initially, who were subsequently shifted to oral medications. Six patients (33.3%) were treated with immunosuppressive therapy – five had received oral azathioprine (50 mg three times a day) and one of them developed deranged liver function test and was shifted to oral mycophenolate. Eight eyes (40%) had recurrence of scleritis and three of them had two recurrences. Five patients developed cataract and only one required surgical intervention. Three patients developed ocular hypertension and all of them responded to antiglaucoma medications. The mean BCVA

Table 1: Clinical profile of patients with posterior scleritis in a tertiary eye care center from Eastern India

Mean age (years)	41.2±10.6 years (26-63)
Duration of follow-up	212.4 days (258-819)
Sex (%)	
Male	8 (44.4)
Female	10 (55.6)
Laterality (%)	
Unilateral	16 (88.9)
Bilateral	2 (11.1)
Presenting complaints (%)	
Diminution of vision	16 (88.8)
Ocular pain	11 (61.1)
Redness	4 (22.2)
Headache	5 (27.8)
Anterior segment signs (number of eyes) (%)	
AC reaction	9 (45)
Anterior scleritis	3 (15)
Eyelid swelling	1 (5)
Posterior segment signs (number of eyes) (%)	
Choroidal/ILM folds	7 (35)
Subretinal fluid at posterior pole	7 (35)
Disc edema	5 (25)
Giant nodular scleritis	1 (5)
Macular star	1 (5)
Laboratory investigations (%)	
Serum ACE	1 (5.6)
Positive IGRA	1 (5.6)
Positive RF	1 (5.6)
Ultrasonography B-scan (number of eyes) (%)	
Thickening of posterior sclera	16 (80)
T-sign	6 (30)
Treatment (%)	
Oral corticosteroid	18 (100)
Intravenous pulse steroid	11 (61.1)
Immunosuppressive	6 (33.3)
Azathioprine	5 (27.8)
Mycophenolate mofetil	1 (5.6)
Recurrence (number of eyes) (%)	8 (40)
Complications (number of eyes) (%)	
Cataract	5 (25)
Ocular hypertension	3 (15)

AC: Anterior chamber, ACE: Angiotensin-converting enzyme, ILM: Internal limiting membrane, RF: Rheumatoid factor, IGRA: Interferon-gamma release assay

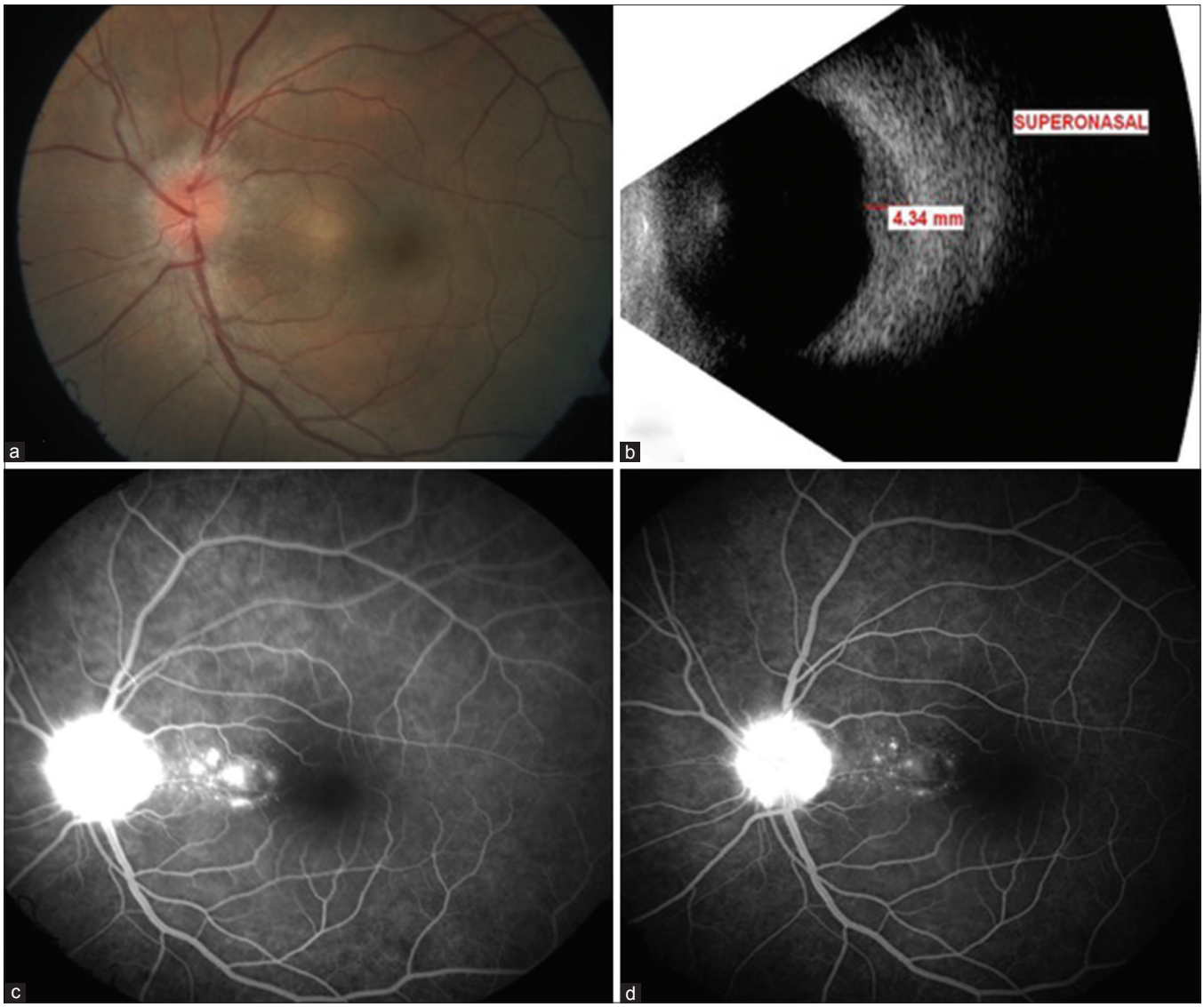

Figure 1: (a) Color fundus photograph of the left eye showing a hyperemic, swollen optic disc with yellowish subretinal mass at posterior pole with overlying subretinal fluid at the posterior pole. (b) Ultrasonography B-scan of the left eye showing localized hyperechoic mass with gross choroidal thickening. (c and d) Fundus fluorescein angiography of the left eye showing pinpoint leaks with pooling corresponding to the lesion in late frame

improved to logMAR 0.06 ± 0.051 at the final follow-up from 0.47 ± 0.45 logMAR at presentation ($P = 0.00608$). On an average, there was a statistically significant improvement of four lines in Snellen chart.

Discussion

Majority of the literature on posterior scleritis from India are isolated case reports. Two large case series on posterior scleritis have been published from India. Twenty years ago, a case series of eight patients of posterior scleritis was published by Biswas *et al*.[4] Recently, a multicenter retrospective study of 114 patients with posterior scleritis by Lavric *et al*.[5] analyzed forty patients from a tertiary eye care center in South India.

The current study showed female preponderance which was in accordance with the other case series on posterior scleritis.[5-8] Compared to other subtypes of scleritis, the posterior scleritis usually occurs in patients with relatively younger age.[3,9] The mean age of our patients in this study (41.2 years) was comparable to similar studies on posterior scleritis[7] and was in accordance with finding of the study by Lavric *et al*.,[5] where a relatively younger age of involvement in posterior scleritis was reported in Indian patients when compared with patients from the United Kingdom.

Ocular pain is an important sign of posterior scleritis and usually results from stretching of nerves that pass through the sclera and swelling of the optic nerve sheath. However, ocular pain is not a constant feature in posterior scleritis, and the disease can occur even in the absence of ocular pain. In a large cohort on posterior scleritis, ocular pain was absent in 36% of the patients with posterior scleritis.[5] In this study, 61.1% of the patients had ocular pain, and the most common symptom complained by the patients was diminution of vision (88.8%).

The current study showed relatively higher number of anterior uveitis (45%). However, anterior uveitis in patients

with posterior scleritis is common and has been reported in approximately 55% of patients in one study.[10] An association with systemic rheumatic disease has been found in 20%–30% of the patients with posterior scleritis.[6-8] Although battery of laboratory tests and thorough systemic evaluation with multidisciplinary approach were done, the current study did not observe any evidence of systemic involvement in patients with posterior scleritis. Although rare, similar observation has been reported in literature.[3] It is not clear whether relatively younger age of the patients (41.2 years) in the current study has any role, who might develop the systemic manifestations of an underlying systemic rheumatic disease at a later age. Furthermore, subtype of scleritis such as necrotizing scleritis is reported to have a high association with systemic rheumatic disease often up to 95% in literature,[7] which was found to have relatively lower systemic association in Indian patients.[11]

Majority of our patients were treated with intravenous pulse corticosteroid therapy, which can be explained by the relatively more aggressive nature of the disease in the current study. Twelve patients (66.7%) had a visual acuity worse than 6/12 (logMAR 0.3) at presentation, and in 88.8% of the patients, the presenting complaint was diminution of vision. Although medications such as immunosuppressives were used less frequently (33.3%), the visual outcome of this study was reasonably good. Aggressive initial treatment with pulse corticosteroid therapy and absence of systemic rheumatic diseases may be attributed to this observation. The presence of an underlying systemic disease has been reported to be associated with visual loss and more severe disease in patients with scleritis.[12] Again, relatively good visual outcomes have been observed in patients of posterior scleritis with lower proportion of associated systemic rheumatologic disease.[3] However, relatively lower use of immunosuppressive agents was reflected in higher incidence of recurrence of posterior scleritis in this study (40%). The absence of any systemic rheumatic disease may be a reason for this lower use of immunosuppressive agents in this study.

Being conducted in a tertiary eye care center, the study does suffer from its own limitation and it will be impractical to ascertain that the current study reflects the true representation of patients with posterior scleritis from Eastern India. However, we believe that this study was able to highlight the clinical pictures of posterior scleritis from Eastern India, many of which are in accordance with the existing literature.

Conclusion

Posterior scleritis is relatively rare but can have vision-threatening presentations. Relatively younger age of presentation, the absence of association with systemic rheumatic disease, relatively higher incidence of anterior uveitis, and the need for aggressive therapy should be kept in mind while managing a patient with posterior scleritis

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AK conceptualized the project and AG collated all the data. AG and AK were directly involved in patient care. PDM wrote the manuscript and was involved in manuscript reviewing and editing. JB was involved in intellectual input along with critical thinking.

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Conflicts of interest

There are no conflicts of interest.

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