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Case Series

Clinical Presentation of Posterior Scleritis in Pediatric Patients: A Case Series from Tertiary Hospital of Western India

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Abstract

Posterior scleritis is an uncommon condition that usually occurs in adults and the diagnosis is often missed. It is even more rare in children. We report three cases of the disease in children. All three patients presented with severe ocular pain. The diagnosis was confirmed on B-scan ultrasonography with positive T-sign. None of the patient had clinical or laboratory evidence of associated systemic disease. All three children required different set of monotherapy or combination therapy for management of the disease. This condition, although rare, should be recognised in children and managed as per the severity of the disease.

Keywords: Retinoblastoma (Rb), Survivors, Families, Quality Of Life (QoL), Health Outcomes, Psychosocial Functioning, Review.

Introduction

Posterior scleritis is an uncommon inflammatory condition that can threaten vision, marked by inflammation of the sclera posterior to ora serrata [1,2]. Its clinical presentation is often nonspecific, posing significant diagnostic challenges owing to its overlapping features with other ocular inflammatory conditions. Unlike other subtypes of scleritis, systemic association with posterior scleritis is relatively rare [2]. The clinical entity is even more uncommon in paediatric age group leading to its often under-diagnosis or misdiagnosis ultimately leading to mistreatment. This article provides a detailed overview of three paediatric patients who were diagnosed with posterior scleritis and managed according to severity of their disease and its recurrence.

Case 1

A 15-years-old female, presented with chief complaint of ocular pain in both eyes for 5 days. Associated complaints were diminution of vision (DOV) since 4 days. Best corrected visual acuity (BCVA) in both eyes were 20/40 and N8 for distance and near, respectively. Anterior segment was quiet in both the eyes. Vitreous did not have any inflammation. Fundus examination of both eyes was within normal limit. Optical coherence tomography (OCT) showed normal macular findings. However, on B-scan ultrasound, T-sign was present bilaterally with raised Retino-choroidal scleral (RCS) thickness. Fundus fluorescein angiography (FFA) revealed multiple pin-point leaks in early phase in the para-papillary area, predominantly over papillo-macular bundle (PMB). Diagnosis of bilateral posterior scleritis was made and patient was started on course of oral steroids as per body weight in tapering manner.

Investigations were done to rule out any systemic associations.

On follow-up after 2 weeks, patient came with all the systemic investigations, which were normal. BCVA improved to 20/20 and N6 in both the eyes, respectively. Bilateral B-scan showed resolution of T-sign and reduction in RCS thickness.

Oral steroids were slowly tapered over the period of 2 months. No recurrence was noted on low dose oral steroids and even after completing the course.

Case 2

A 14-years-old male came with chief complaint of bilateral ocular pain for 3 days. Associated complaints were redness in both the eyes. BCVA at presentation was 20/30 and N8 for distance and near in both the eyes. Bilateral conjunctival hyperemia with engorged vessels showed associated anterior scleritis (Figure 1 E) Anterior segment, vitreous and fundus findings were within normal limit. FFA could not be done as parents did not agree for it. B-scan of both eyes revealed raised RCS thickness along with bilateral T-sign. Patient was started on Oral NSAIDs and systemic investigations were done. On follow-up after 2 weeks, the BCVA improved to 20/20 and N6 in both eyes. T-sign had resolved on B-scan in both the eyes along with reduction in RCS thickness. No systemic association was seen on blood investigations. No recurrence was noted up to 52 weeks post resolution.

Case 3

A 16-years -old female patient came with chief complaints of bilateral ocular pain with diminution of vision for 10 days. BCVA in both eyes were Finger counting 1 meter. Anterior segment and vitreous findings were normal. Fundus revealed sub-retinal fluid pockets (Figure 1A) over posterior pole and inferior exudative retinal detachment. OCT revealed pockets of neurosensory detachment (Figure 1C) with choroidal folds. Bilateral B-scan showed T-sign (Figure 1D) with inferior exudative retinal detachment. FFA revealed multiple pin-point leakage (Figure 1B) over posterior pole and in para-papillary region in early phase. Systemic investigations were advised. Patient was started on intravenous methyl-prednisolone 500 mg for 3 consecutive days followed by oral steroids as per body weight along with oral NSAIDs. On 1 week follow-up, patients' BCVA improved to 20/200 in both eyes and systemic investigations were within normal limit. OCT showed resolving fluid pockets and exudative RD also started to settle down on follow-up, clinically. Systemic steroids were then tapered accordingly. After systemic steroids were tapered, patient had recurrence in form of subretinal fluid pockets over posterior pole. A high dose of oral steroids as per body weight were started again and this time patient was started on tablet Azathioprine for the same.

Discussion

Posterior scleritis refers to inflammation of the sclera posterior to the ora serrata and is estimated to account for only 2% to 12% of all cases [1,3]. Incidence of posterior scleritis in children is even rarer than in the adult population and is often idiopathic with no associated systemic disease, or concurrent anterior segment inflammation or pain and occurs both in males and females. However, it represents a larger portion of the overall incidence of scleritis in this age group [3-5].

Majumder., *et al*. reported pain as being the most common presenting complaint in paediatric scleritis (90%), followed by redness in 60% of patients [4]. Painless posterior scleritis has been reported previously [6], but despite that, pain remains the hallmark presenting symptom in scleritis and its recognition is crucial for early diagnosis and appropriate treatment.

Ophthalmic ultrasound (B-scan) plays a vital role in confirming the diagnosis and evaluating the extent of inflammation in posterior scleritis. It allows the visualization of the thickened scleral and the retrobulbar edema and fluid collection, which is the pathognomonic T-sign in posterior scleritis [5,7,8]. Cheung and Chee reported in one of the largest case series of paediatric patients with posterior scleritis, where all eyes (100%) demonstrated positive T-sign on B-scan ultrasound [3]. It is a non-invasive and cost-effective modality that can be particularly beneficial in paediatric patients, aiding in early accurate diagnosis. As per Cheug., *et al*. [3] and Majumder., *et al*. [4] most of the patient of posterior scleritis does not have any systemic association. Similar finding is noted in our case

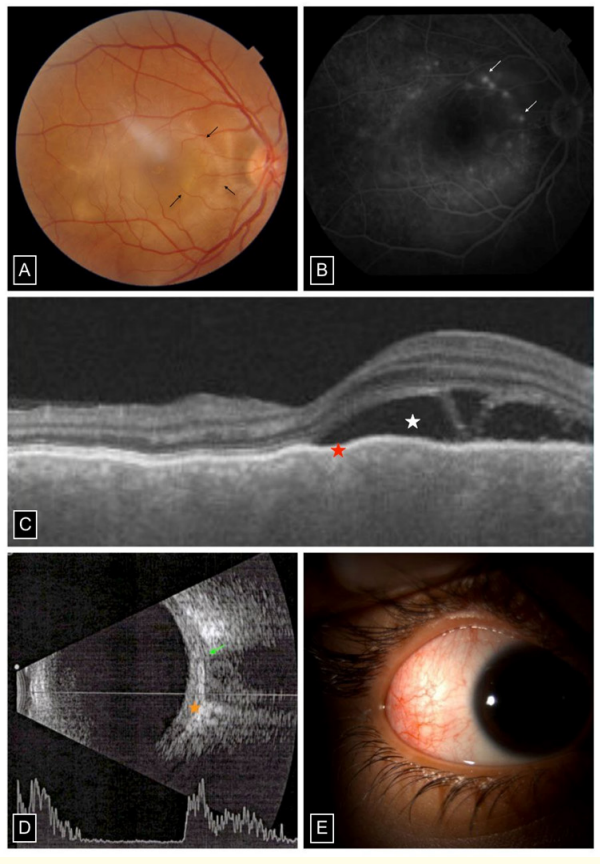

Figure 1: A, Fundus photo right eye showing multiple pockets of exudative retinal detachment (black arrows); B, FFA showing multiple pin-point hyperfluorescent leaks (white arrows); C, OCT shows neurosensory detachment (white star) along with choroidal folds (red star); D, B-scan showing increased retino-choroidal thickness (orange star) along with T-sign (green star); E, Slit-lamp photo showing conjunctival hyperaemia with engorged vessels showing associated anterior scleritis.

series where none of the patient had any systemic association with posterior scleritis. However, in the above-mentioned studies, most of the children had unilateral involvement; while our case series had both eyes involvement in all 3 children. Also in the aforementioned studies, children received IMT for relapse or recurrence. While in our series, two patients did not require the need of IMT, while in one patient IMT was required for relapse of the disease.

Conclusion

Posterior scleritis is often misdiagnosed and mistreated. The rarity and variable presentation of posterior scleritis makes it difficult to diagnose, and it is often misdiagnosed. Primary therapy includes oral NSAIDS alone, systemic corticosteroids alone, or a combination of both. Approximately one-third of patients require immune-modulator therapy which particularly helps in manging recurrences.

Given its varied presentation, clinicians must have a high index of suspicion for posterior scleritis when presentation is atypical for a more common pattern of ocular inflammation.

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Conflict of Interest

The authors declare no conflicts of interest.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. The patient provided consent for his/her image and other clinical information to be reported in the journal. The patient understands that their names and initials will not be published, and due efforts will be made to conceal their identity; however, anonymity cannot be guaranteed.

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