Posterior Scleritis Secondary to Sarcoidosis

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Abstract
Purpose: To report a case of posterior scleritis presumed secondary to sarcoidosis.
Methods: Case presentation.
Results: A 79-year-old male with a history of sarcoidosis presented with asymptomatic conjunctival injection and chemosis of the right eye. The patient presumptively was diagnosed with anterior scleritis of the right eye. At a follow-up visit, examination and diagnostic imaging revealed choroidal folds and increased choroidal thickness. He was diagnosed with posterior scleritis presumed secondary to sarcoidosis in the right eye. The patient was started on topical and oral steroids and the choroidal thickness and folds improved at one-week follow-up.
Conclusion: Posterior scleritis is an unusual manifestation of sarcoidosis, but it should be considered in the differential diagnosis of patients with sarcoidosis who present with a red, painful eye, and choroidal folds and thickening.

Key words: posterior scleritis, choroidal folds, sarcoidosis

Introduction:
Posterior scleritis is a rare but potentially vision-threatening condition that is often misdiagnosed due to its variable clinical presentation. Posterior scleritis represents 2-12% of all cases of scleritis, half of which can be associated with systemic disease, most commonly autoimmune inflammatory conditions. Posterior scleritis is uncommonly associated with sarcoidosis. Herein we present a case of posterior scleritis in a patient with sarcoidosis and review its diagnosis and management.

Case Presentation:
A 79-year-old male reported to the eye clinic with a complaint of a red and swollen right eye for two days. He denied symptoms of itchiness, pain, or photophobia. Ocular history was remarkable for an isolated episode of anterior scleritis in the left eye one year prior to presentation. His medical history was positive for pulmonary sarcoidosis diagnosed in 2015, hypertension, and hyperlipidemia. His medications included atorvastatin, atenolol, lisinopril, and apixaban. He was not currently being treated for sarcoidosis.

Upon examination, the patient’s best-corrected visual acuity was 20/40 for both the right eye and left eye. Finger counting fields revealed a superior field constriction in the right eye with full fields in the left eye. Pupils and extraocular motilities were unremarkable. An anterior segment exam showed upper lid inflammation with a mild ptosis, moderate conjunctival/scleral injection and chemosis, trace corneal microcystic edema adjacent to the nasal limbus, and an anterior chamber reaction of 1+ cells with no flare in the right eye (Figure 1). The left eye was unremarkable. There was a clear and centered PCIOL in the right eye and a mild nuclear sclerotic cataract in the left eye.

Intraocular pressures with Goldmann applanation tonometry were 18mmHg in the right eye and 19mmHg in the left eye.

Undilated examination of the posterior pole was unremarkable in both eyes.

The patient was diagnosed with non-granulomatous anterior uveitis of the right eye with a suspected anterior scleritis. The gross superior visual field constriction can be attributed to the edematous right upper eyelid. The patient was prescribed topical prednisolone acetate 1% every four hours while awake for the right eye, and to follow-up the next day.

Figure 1A-B. Initial anterior segment presentation of the right eye; moderate conjunctival/scleral injection and chemosis.
At the one-day follow-up the patient reported mild improvement of the right upper lid inflammation, and minimal improvement in conjunctival/scleral injection and chemosis. The anterior chamber reaction was improved to trace cells. Intraocular pressure was 9mmHg in the right eye. Dilated fundus examination was unremarkable. The patient was instructed to continue topical prednisolone acetate 1% to the right eye every four hours while awake and a referral to ophthalmology service was arranged for the next day.

In the ophthalmology clinic, the patient reported his symptoms had worsened. The right upper lid inflammation had returned and there was moderate conjunctival/scleral injection and chemosis, along with moderate amount of microcystic edema adjacent to the nasal limbus. The anterior chamber was deep and quiet. Topical administration of phenylephrine 2.5% resulted in no blanching of the engorged vessels, and fundus examination remained unremarkable. The patient was diagnosed with diffuse anterior scleritis of the right eye. Oral prednisone 50mg daily, and oral pantoprazole 20mg daily for gastrointestinal prophylaxis. The patient was instructed to discontinue prednisolone acetate 1% and to return to clinic in four days for follow-up.

Laboratory work-up including complete blood count, rheumatoid factor, antinuclear antibody, angiotensin-converting enzyme, antineutrophil cytoplasmic antibodies, rapid plasma regains, fluorescent treponemal antibody absorption, and compliment component 3 and component 4 were unremarkable. Erythrocyte sedimentation rate results were abnormally elevated at 87 mm/hour, and a T-spot testing was ordered but not obtained.

When the patient returned for follow-up four days later, there was noted improvement of the right upper lid inflammation. The conjunctival/scleral injection was mild to moderate, and the corneal microcystic edema was improving. Dilated fundus examination revealed presence of a choroidal effusion in the temporal periphery of the right eye (Figure 2). Macular ocular coherence tomography (OCT) showed RPE undulation consistent with choroidal folds (Figure 3). The posterior sclera and choroid were noted to be thickened with B-scan ultrasonography. A faint "T-sign" was also present consistent with fluid in the subtenon space (Figure 4). Fundus fluorescein angiography (FFA) showed no delays in choroidal, arterial, or venous filling. No signs of optic disc leakage or staining of retinal vasculature was present (Figure 5A-B).
The diagnosis was revised to posterior scleritis, likely secondary to sarcoidosis. The patient was instructed to continue the oral steroid and start use of topical difluprednate 0.05% four times daily, and topical atropine sulfate 1% two times daily and to follow-up in one week.

The patient returned for follow-up one week later with improvement in vision of the right eye to 20/25, significant improvement of the anterior segment findings and resolution of the choroidal effusion (Figure 6 & 7). Prednisone was decreased to 40mg daily with a 5mg per week taper. The patient’s primary care physician was alerted of the patient’s ocular findings and a consultation was placed for further evaluation with rheumatology.

Discussion:
This report highlights a case of posterior scleritis presumed secondary to sarcoidosis. While it presented initially as an anterior scleritis, the subsequent development of a choroidal effusion and folds heralded the development of a posterior scleritis. The patient responded well to a course of oral steroids.

Scleritis
Scleritis is a severe ocular inflammatory condition that can present unilaterally or bilaterally and affects the anterior and/or posterior sclera. Scleritis can be idiopathic, infectious, or non-infectious. The pathophysiology of scleritis is unknown but typically occurs more often in persons with autoimmune inflammatory conditions, most commonly rheumatoid arthritis. Scleritis is not often associated with sarcoidosis but should be considered as a differential diagnosis when an episode occurs.

Ocular involvement occurs in approximately 20-30% of patients with sarcoidosis and may be the only presenting symptom in about 5% of these patients. Sarcoidosis-related scleritis is an uncommon ocular manifestation, but may present as anterior diffuse, anterior nodular, or posterior scleritis. Diffuse anterior scleritis is generally the most common inflammatory presentation. The less frequent presentation, posterior scleritis, makes up 2-12% of all scleritis cases.

The hallmark presenting symptom of posterior scleritis is moderate to severe deep, dull, boring pain. In addition, there may be associated pain on extraocular motility, proptosis, decreased vision, and an associated anterior scleritis. However, it is not unusual for a patient to present with absence of any ocular pain, as was the case with our patient.

Clinical signs of posterior scleritis are variable and may include eyelid edema, ciliary or conjunctival congestion, anterior uveitis, corneal edema, shallowing of the anterior chamber, limitation of extraocular motility, retinal striae, disc swelling, serous retinal detachment, choroidal folds, and/or choroidal effusions.

Ocular Diagnostic Testing
Due to the variability of its clinical presentation, diagnostic imaging plays a vital role in diagnosing those with suspected posterior scleritis (Table 2).

Systemic Workup
All patients that present with posterior scleritis should have a systemic workup to identify the cause of the condition and any associated metabolic condition (Table 3).

Treatment/management of posterior scleritis
Posterior scleritis can be challenging to diagnose and requires immediate therapy. The first line of treatment can be oral NSAIDs such as ibuprofen 600-800mg QID, piroxicam 20mg QD, or naproxen 375mg BID. Oral corticosteroids can also be used initially or in cases where treatment with oral NSAIDs are ineffective. The typical treatment includes prednisone 1mg/kg/day or approximately 60-80mg daily. If the condition is unresponsive to corticosteroids, the next step involves immunosuppressive/antimetabolite agents such as methotrexate, azathioprine and mycophenolate. However, these agents are not fast-acting and could take up to six months to fully take effect. In cases where patients require a more rapid solution, biologic tumor necrosis factor (TNF) inhibitors such
as rituximab or infliximab are initiated.\textsuperscript{1,2,6,13} Prompt diagnosis, appropriate, and aggressive treatment are key to quick recovery and preservation of vision.\textsuperscript{1}

**Conclusion:**
Posterior scleritis is a rare but potentially vision threatening ocular condition. Prompt diagnosis and collaboration with primary care and, as needed, subspecialty physicians are essential to the management of patients with posterior scleritis.

**References:**