Posterior scleritis-induced optic neuropathy and exudative retinal detachment – A challenging diagnostic dilemma

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We report three cases of posterior scleritis (PS) to analyze the clinical profile and ultrasonographic and fluorescein angiography features of this rare disorder. Fundus findings included serous retinal detachment (RD), disc edema, disc hyperemia, corkscrewed retinal vessel, and retinal folds. Ultrasonography revealed a variable degree of thickening of the posterior eye wall (choroid and sclera). Fluorescein angiography revealed persistent dye leakage from the disc and early pinpoint areas of hyperfluorescence with pooling of dye in late frames of an angiogram. Optical coherence tomography showed serous macular detachment in all cases at the time of presentation. The purpose of this manuscript was to describe three cases of PS associated with optic neuropathy and exudative RD previously misdiagnosed with a range of conditions. This case study also demonstrates the importance of B-scan ultrasonography and fluorescein angiography for the appropriate diagnosis of PS and also the effectiveness of systemic corticosteroid therapy.

Key words: B-scan ultrasonography, exudative retinal detachment, fundus fluorescein angiography, optic neuropathy, posterior scleritis

Posterior scleritis (PS) is an uncommon entity, which refers to an inflammation of the sclera posterior to the ora serrata that is severe enough to cause abnormalities in the posterior segment (choroid, retina, and optic nerve) and the anterior segment of the eye.¹,² The classic symptoms associated with PS are the periorcular pain, pain on movement, decreased vision, and redness of the eye, but it must be stressed that many patients have none or only one of these. PS has been reported to account for only 2%–12% of all cases of scleritis and in up to 50% of cases, it can be bilateral.³ However, its actual incidence is probably underrecognized due to its clinical features (papillitis, disc edema, retinal folds, choroidal folds, serous retinal detachment [RD], etc.) that may resemble other intra- or extraocular inflammatory and neoplastic conditions.⁴ Middle-aged women are more frequently affected than men.⁵ Ultrasonography is the key investigation necessary to make a diagnosis of PS, and computed tomography (CT) scan, magnetic resonance imaging (MRI), and fundus fluorescein angiography (FFA) can also be used as ancillary tests.⁶

Herein, we describe three cases of PS associated with optic neuropathy and exudative RD previously misdiagnosed with a range of conditions and the role of ultrasound and FFA findings in the diagnosis and also demonstrate the effectiveness of high-dose methylprednisolone therapy.

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Submission: 05.03.2019; Decision: 10.10.2019
Acceptance: 22.10.2019; Web Publication: 06.03.2020

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Cite this article as: Bawankar P, Das D, Bhattacharjee H, Soibam R. Posterior scleritis-induced optic neuropathy and exudative retinal detachment – A challenging diagnostic dilemma. J Clin Ophthalmol Res 2020;8:39-42.
Case Report

We describe three cases which were diagnosed as having PS in our hospital, which is a referral eye center. The information regarding age, sex, laterality, the preliminary diagnosis of the referring ophthalmologist, pre- and posttreatment best-corrected visual acuity, and the treatment advised were noted [Table 1]. Status of retinal vessels, optic nerve head, secondary RD, retinal or choroidal folds, etc., was revealed on

Figure 1: Case 1 – (a) Color fundus image showing hyperemic edematous optic disc, exudative retinal detachment at the posterior pole, retinal folds, and corkscrewed retinal vessels. (b-d) Early, mid, and late phases of the angiogram showing early hyperfluorescence with late pooling of dye in the subretinal space and persistent dye leakage from the disc

Figure 2: Case 1 – (a) Color fundus image showing resolution of disc edema, macular detachment, and retinal folds after 2 months of treatment. (b) Pretreatment optical coherence tomography image showing subretinal fluid at the macula. (c) Posttreatment optical coherence tomography image showing resolution of exudation and flattening at the macula. (d and e) B-scan ultrasonography showing choroidal and scleral thickening with exudative detachment at the macula

Figure 3: Case 2 – (a) Fundus image showing exudative macular detachment which appears like central serous chorioretinopathy. (b) Optical coherence tomography image showing focal detachment at the macula. (c) B-scan ultrasonography showing classic “T”-sign and marked thickening of the posterior eye wall. (d) Late phase of angiogram showing persistent dye leakage from the disc and in the subretinal space at the macula

Figure 4: Case 3 – (a) Color fundus image showing hyperemia with disc edema, subretinal fluid accumulation at the macula. (b) Optical coherence tomography image showing subretinal fluid at the macula. (c) B-scan ultrasonography showing gross choroidal and scleral thickening. Fluorescein angiography showing (d) early pinpoint hyperfluorescence in the early phase and (e) areas of late pooling of dye and diffuse disc leakage in the late phase of angiogram
Hyperemia with disc edema, systemic steroid treatment 6 months. Age/sex OD Hyperemic optic disc with blurred vision in OD. 6 months. Pseudotumor Hyperemia with disc edema and choroidal folds, one should consider orbital tumor, inflammatory pseudotumor, or thyroid ophthalmopathy in the differential diagnosis. In this sense, PS is often misdiagnosed by the referring ophthalmologist who may refer these patients to other physicians rather than ophthalmologists, particularly if ultrasonography findings are nonspecific or unremarkable. However, it is unable to detect some early forms of this disease, and a characteristic T-sign may be absent in up to 75% of PS cases. In this sense, PS is often an underrecognized form of inflammation most likely due to its location and associated nonspecific signs and symptoms. A high index of suspicion is usually required for the diagnosis, and it is usually based on a compatible clinical spectrum in addition to indirect signs observed on BSU and other ancillary tests such as FFA, CT, and MRI. Ultrasonography is the most helpful ancillary test in the diagnosis of PS. However, it is unable to detect some early forms of this disease, and a characteristic T-sign may be absent in up to 75% of PS cases.

**Table 1: Clinical profile, referral diagnosis, and the treatment characteristics of three cases of posterior scleritis**

| Case number | Age/sex | Laterality | Pretreatment VA | Referral diagnosis | Treatment | Posttreatment VA | Follow-up period |
|-------------|---------|------------|-----------------|--------------------|-----------|-----------------|-----------------|
| 1           | 28/male | OD         | 6/12 N12        | Pseudotumor        | Systemic steroid | 6/6 N6          | 5 months        |
| 2           | 26/female | OD         | 6/9 N8          | CSCR               | Systemic steroid | 6/6 N6          | 8 months        |
| 3           | 34/male | OD         | 6/12 N8         | Papillitis         | Systemic steroid | 6/6 N6          | 6 months        |

OD: Oculus dexter; CSCR: Central serous chorioretinopathy; VA: Visual acuity

**Table 2: Imaging characteristics of three cases of posterior scleritis**

| Case number | Fundus findings | Ultrasonographic features | OCT | FFA |
|-------------|-----------------|---------------------------|-----|-----|
| 1           | Hyperemia with disc edema, corkscrewed retinal vessel, retinal folds, and exudative detachment at the posterior pole [Figure 1a]. Post-treatment color fundus image [Figure 2a] | Marked choroidal and scleral thickening (3.8 mm); ERD at the posterior pole; peribulbar fluid accumulation [Figure 2d and e] | Neurosensory detachment at the macula [Figure 2b]. Post-treatment OCT image [Figure 2c] | Persistent dye leakage from the disc with pooling of dye in the subretinal space of the right eye. [Figure 1b, c, and d] |
| 2           | Hyperemic optic disc with blurred margin; subretinal fluid at the macula [Figure 3a] | Marked thickening of the posterior eye wall: “T.” sign present [Figure 3c] | Exudative detachment at the macula [Figure 3b] | Pinpoint areas of leakage in the early phase of angiogram with persistent dye leakage from the disc [Figure d] |
| 3           | Hyperemia with disc edema and subretinal fluid accumulation at the macula [Figure 4a] | Choroidal and scleral thickening (2.5 mm); elevation of the optic nerve head; no “T.” sign [Figure 4c] | Few intraretinal cystic spaces with subretinal fluid [Figure 4b] | Early pinpoint areas of hyperfluorescence with late pooling of dye; disk leakage [Figure 4d and e] |

ERD: Exudative retinal detachment, OCT: Optical coherence tomography, FFA: Fundus fluorescein angiography
choroidal melanoma, metastatic carcinoma to the choroid, or choroidal hemangioma should be excluded.\textsuperscript{[7]} In cases with a serous detachment of the retina, conditions such as uveal effusion syndrome, Vogt–Koyanagi–Harada disease, or central serous retinopathy should be kept in mind.\textsuperscript{[8]}

In case 1, moderate-to-severe pain, pain on eye movement, conjunctival chemosis, proptosis, and disc edema indicated a diagnosis of the pseudotumor. However, associated anterior scleritis and anterior uveitis indicated a suspicion of PS. Pseudotumor can closely mimic an acute PS, particularly when associated with anterior scleritis. Both conditions can cause retino-choroidal striae, scleral thickening, retrobulbar edema, and extraocular muscle enlargement.\textsuperscript{[9]} However, in PS, intraocular findings such as anterior uveitis and retinal and optic nerve involvement are more prominent than extraocular findings. The diagnosis of pseudotumor often depends on mostly demonstration of an orbital mass by ultrasonography or CT scan;\textsuperscript{[9]} therefore, PS was the distinct possibility in this case.

In case 2, a diagnosis of central serous chorioretinopathy (CSR) was made based on the serous RD involving the macular area. CSR does not cause pain and has clear subretinal fluid. FFA and ultrasonography can certainly differentiate PS from CSR. CSR nearly always has only a single leak on FFA and on ultrasonography shows only a serous RD, not the choroidal and scleral thickening, disc edema, retrobulbar edema, and “T-” sign that are seen in PS.

PS may be associated with optic disc swelling in up to 17% of patients.\textsuperscript{[4]} In case 3, mild-to-moderate pain associated with defective vision, disc edema, exudative macular detachment, and characteristic findings on USG and FFA supported the diagnosis of PS.

The management of PS is less challenging in contrast to the diagnosis. It is a treatable disease, as shown by the prompt remission of signs and symptoms in all patients we treated with systemic corticosteroids. To conclude, a high index of suspicion should be kept in any disc edema associated with exudative RD involving the posterior pole if associated with pain. In such a case, a BSU and an FFA can be done to establish the diagnosis. Ophthalmologists must remain aware of the protean manifestations of this unique entity, and early diagnosis of PS is most important due to its excellent response to anti-inflammatory medications, particularly with systemic steroids.

Acknowledgment
The authors would like to acknowledge Sri Kanchi Sankara Health and Educational Foundation, Guwahati, Assam, India.

Financial support and sponsorship
This study was financially supported by Sri Kanchi Sankara Health and Educational Foundation.

Conflicts of interest
There are no conflicts of interest.

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