An Atypical Presentation of Posterior Scleritis
A Ramanathan, A Gaur

Citation

Abstract
We report a case of an unusual presentation of posterior scleritis. A 13 year old girl presented with progressive monocular visual loss over one week. She did not complain of pain as a significant symptom. Examination of the fundus revealed a pale raised subretinal lesion temporal to the fovea with overlying exudative retinal detachment. A B-scan ultrasound confirmed choroidal and scleral thickening, retinal detachment and fluid in the sub-Tenon’s space. Fundus fluorescein angiography showed multiple areas of progressively increasing hyperfluorescence (leakage of dye) temporal to the fovea. Systemic evaluation of this patient did not reveal an underlying systemic disorder. The patient was commenced on high dose oral corticosteroids which resulted in resolution of symptoms and clinical features. She made full visual recovery after six weeks of treatment. This case illustrates an interesting presentation of posterior scleritis in a 13 year old and explores clinical features, treatment and prognosis in this age group.

CASE REPORT
A 13 year old girl presented with a 1 week history of progressive visual loss affecting her left eye with an associated intermittent dull ache around the eye. She did not give a history of a preceding flu-like illness or trauma to the eye. She had no significant past medical or ocular history. Best corrected visual acuity in the left eye was counting fingers at 1 meter and 6/6 in the right eye. Examination on the anterior segment was unremarkable in both eyes. In particular, there was no evidence of proptosis or painful restriction of eye movements.

Dilated fundoscopy of the left eye revealed a pale subretinal raised lesion temporal to the fovea with overlying exudative retinal detachment involving the macula (figure 1a). B-scan ultrasonography showed diffuse thickening of the choroid with low acoustic reflectivity. The sclera showed thickening and there was evidence of fluid in the sub-Tenon’s space (figure 1b).
Figure 2
Figure 1b: Left B-scan ultrasound shows diffuse thickening of the choroid with low acoustic reflectivity. There is scleral thickening and evidence of fluid in the sub-Tenon’s space.

Fundus fluorescein angiography showed multiple areas of progressively increasing hyperfluorescence temporal to the fovea with pooling of dye within the exudative retinal detachment (figure 2a-c).

Figure 3
Figure 2a: Left fluorescein angiography (FFA) showing the arteriovenous phase at 23 seconds with minimal hyperfluorescence infero-temporal to the fovea.

Figure 4
Figure 2b: Left FFA at 37.6 seconds shows multiple foci of hyperfluorescence progressively increasing in the later frames (figure 2c). Figure 2c also shows increased hyperfluorescence in the overlying area of retinal detachment.

Figure 5
Figure 2c: FFA at 1.02.7 minutes.

General physical examination was unremarkable. Further investigations including full blood count, ESR, CRP, Rheumatoid factor, ANA, ANCA, ACE were within normal range. Serological tests for Toxoplasmosis, Lyme disease and Syphilis were negative. In addition, urinalysis and chest x-ray showed no abnormalities.

The patient was commenced on 1mg/ kg/ day Prednisolone, starting at 40 mg which was tapered to 35 mg and 30mg at 1 week intervals. After 2 weeks of systemic steroid treatment, visual acuity improved to 6/18. Fundoscopy revealed
resolution of the retinal detachment, pigment deposition at
the macula and a small area of residual inflammation
temporal to the fovea (figure 3a). B-scan ultrasonography
showed resolution of the retinal detachment and sub-Tenon’s
fluid (figure 3b). The steroids were gradually tapered down
at 5mg per week and her vision improved to 6/6 after six
weeks of treatment.

**Figure 6**
Figure 3a: Left colour fundus showing pigment deposition at
the macula, a small area of residual inflammation temporal
to the macula and re-attached retina.

Figure 7
Figure 3b: Left B-scan ultrasound showing resolution of the
retinal detachment and sub-Tenon’s fluid.

**DISCUSSION**

On initial presentation, fundus examination revealed an
exudative retinal detachment together with a pale whitish
raised subretinal lesion. A preliminary diagnosis of a
haemangioma causing an exudative retinal detachment was
made. There were no ocular clinical features such as painful
eye movement or positive anterior segment findings to point
towards an inflammatory cause. The B-scan ultrasound
imaging proved to be a vital diagnostic tool in this case. It
demonstrated choroidal and scleral thickening and the T-
sign, diagnostic of posterior scleritis. Thickening of the
choroid with low acoustic reflectivity excluded a choroidal
haemangioma. Another relevant differential diagnosis in this
case would be Acute Multifocal Placoid Pigment
Epitheliopathy (AMPPE). However, the clinical features did
not include a history of prodromal symptoms and the
fluorescein angiographic findings did not show areas of
choroidal hypofluorescence characteristic of AMPPE.

Based on clinical features of a dull intermittent eye pain and
ultrasound findings, a diagnosis of posterior scleritis was
made. The patient was given a course of systemic steroids
after which she made a full clinical recovery. Further
investigations did not find an underlying associated
connective tissue disorder.

Posterior scleritis is a relatively uncommon inflammatory
disorder, frequently associated with systemic autoimmune
diseases and has the potential to cause irreversible visual
loss if left untreated. Studies have highlighted the varied
clinical presentation of posterior scleritis and its ability to
mimic other disorders such as orbital lymphoma or
choroidal melanoma. Pain has been documented as a
cardinal and commonly predominant feature of posterior
scleritis. However, in this case, the patient complained of
only a mild intermittent ache around the eye. Previous cases
have been reported where pain has been absent in the
presentation of posterior scleritis which, in turn, high-
lights the importance of B-scan ultrasound as a diagnostic
investigation for posterior scleritis. Interestingly, an
underlying systemic disorder was not found in these reports
of an 18 and 21 year old with posterior scleritis as with the
case reported here and clinical abnormalities resolved with
oral prednisolone treatment. It has been previously noted
that cases of posterior scleritis in adolescents do not appear
to be associated with an underlying systemic disease in
contrast to the adult variant. Posterior scleritis in children,
even in its severe form, has a good visual prognosis with the
commencement of early appropriate treatment. Patients
over the age of 50 years with posterior scleritis are more
likely to experience sight loss and have an associated
systemic disease which requires aggressive
immunosuppressive treatment.

Treatment options for posterior scleritis include non-
steroidal anti-inflammatory drugs and oral corticosteroids.
Patients who do not respond to these treatments are offered
An Atypical Presentation of Posterior Scleritis

immunosuppressive therapy such as Methotrexate or Azathioprine. In refractory cases, immunomodulatory drugs such as Infliximab may be used.

This case illustrates that posterior scleritis can present with reduced vision without pain as a predominant feature. As it is a rare treatable condition with potentially serious consequences, a high index of suspicion is necessary to make the diagnosis in unusually presenting cases as with this young 13 year old girl. The report also demonstrates the pathognomic features of posterior scleritis on B-scan ultrasound.

References
An Atypical Presentation of Posterior Scleritis

Author Information
Asha Ramanathan, MBBS, BSc
Department of Ophthalmology, Royal Glamorgan Hospital

Amit Gaur, MBBS, MD, FRCS
Consultant Ophthalmologist, Royal Glamorgan Hospital