

An Atypical Presentation of Posterior Scleritis

A Ramanathan, A Gaur

Citation

A Ramanathan, A Gaur. *An Atypical Presentation of Posterior Scleritis*. The Internet Journal of Ophthalmology and Visual Science. 2009 Volume 8 Number 2.

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 funduscopy 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 1

Figure 1a: Left colour fundus showing a pale subretinal lesion temporal to the fovea with surrounding exudative retinal detachment involving the macula.

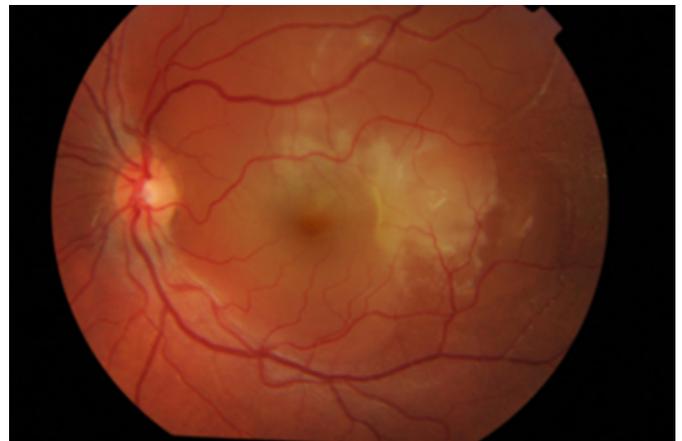
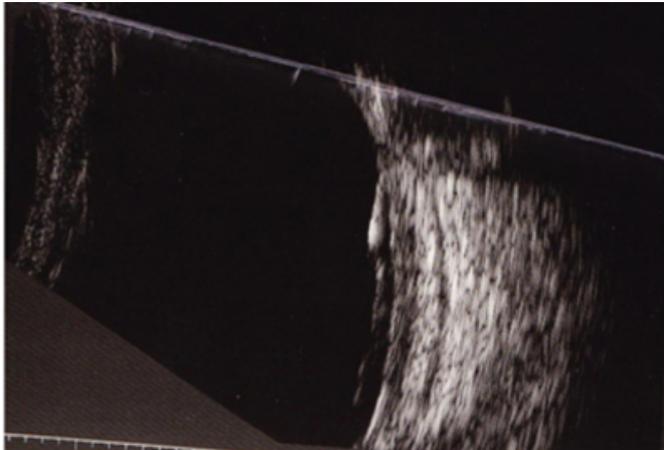


Figure 2

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.

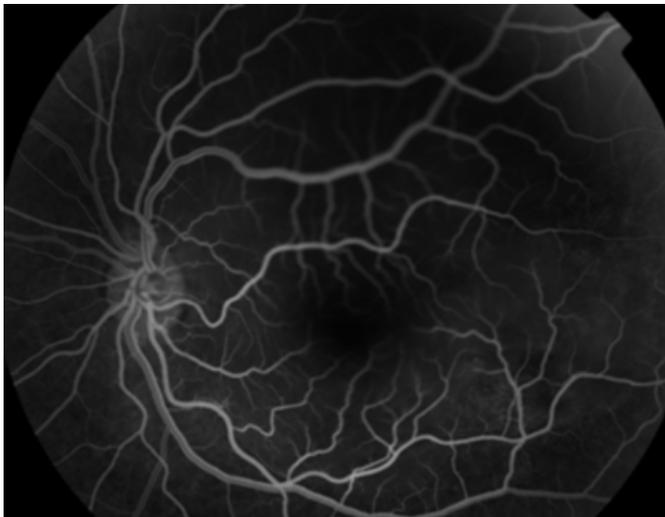


Figure 4

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.

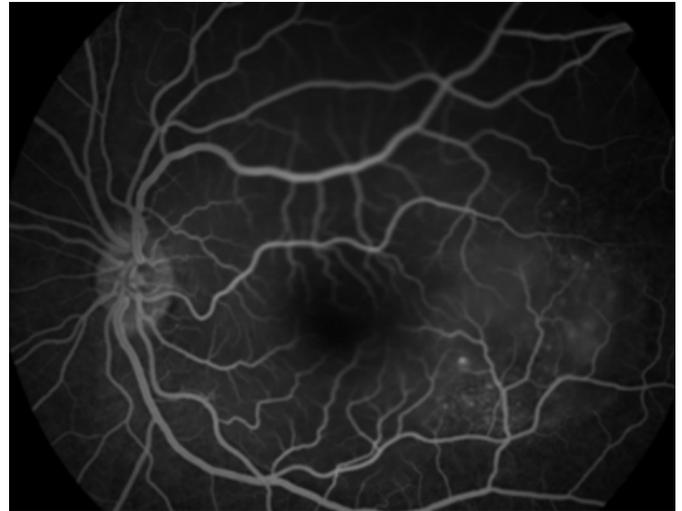
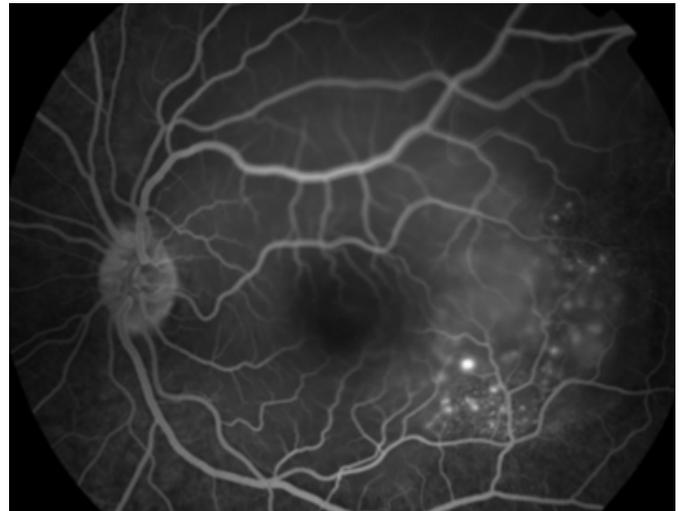


Figure 5

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.

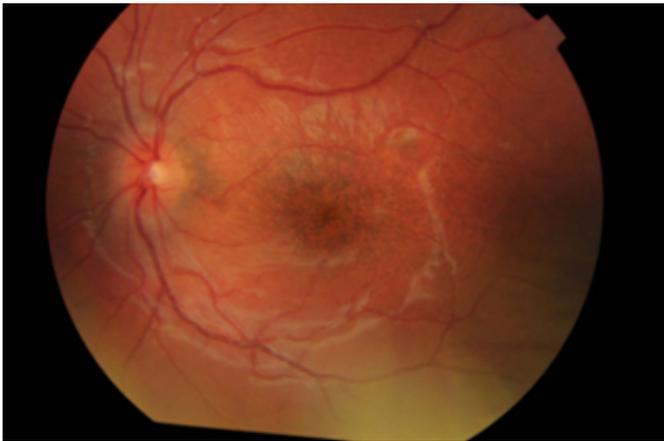
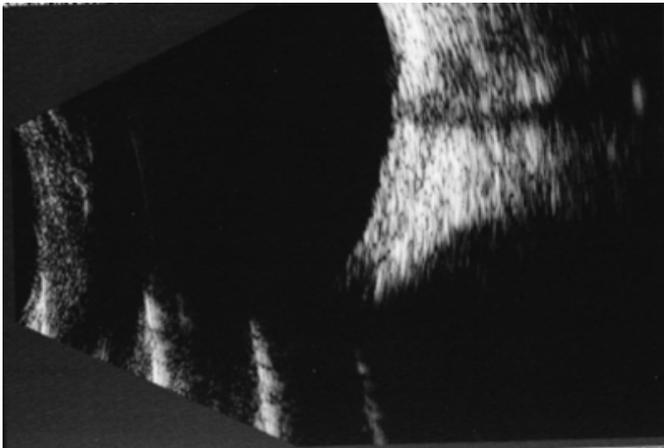


Figure 7

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.¹ Studies have highlighted the varied clinical presentation of posterior scleritis² and its ability to mimic other disorders such as orbital lymphoma³ or choroidal melanoma.⁴ Pain has been documented as a cardinal and commonly predominant feature of posterior scleritis.² 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^{5,6,7} which, in turn, highlight 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^{6,5} 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.⁸ Posterior scleritis in children, even in its severe form, has a good visual prognosis with the commencement of early appropriate treatment.^{8,9} 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.¹

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

immunosuppressive therapy such as Methotrexate or Azothioprine.¹⁰ In refractory cases, immunomodulatory drugs such as Infliximab may be used.¹¹

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

1. McCluskey, Watson PJ, Lightman PG: Posterior scleritis: clinical features, systemic associations, and outcome in a large series of patients. *Ophthalmology*. 1999; 106(12):2380-6.
2. Biswas J, Mittal S, Ganesh SK, Shetty NS: Posterior scleritis: clinical profile and imaging characteristics. *Indian J. Ophthalmology*. 1998; 46 (4):195-202.
3. Dorey SE, Clark BJ, Christopoulos VA, Lightman S: Orbital lymphoma misdiagnosed as scleritis. *Ophthalmology*. 2002; 109(12):2347-50.
4. Demirci H, Shields CL, Honavar SG, Shields JA, Bardenstein DS: Long-term follow-up of giant nodular posterior scleritis simulating choroidal melanoma. *Archives of Ophthalmology*. 2000; 118(9):1290-2.
5. Dodd EM, Irarrazaval LA: Bilateral Posterior scleritis. *Ocul Immunol Inflamm*. 1997; 5(4):267-9.
6. Saika P, Nashed AH, Helbig H, Hillenkamp J: Bilateral posterior scleritis: An idiopathic Painless presentation. *Ocul Immunol Inflamm*. 2010; Oct
7. Slagle WS, Boothe KH, Musick AN, Eckermann DR, Slagle AM: Idiopathic orbital inflammatory syndrome without pain: a case report. 2010; 81(3):146-52.
8. Wald KJ, Spaide R, Patalano VJ, Sugin S, Yannuzzi LA: Posterior scleritis in children. *Am J Ophthalmol*. 1992; 15; 113(3):281-6.
9. Woon WH, Stanford MR, Graham EM: Severe idiopathic posterior scleritis in children. *Eye (Lond)*. 1995;9 (Pt 5):570-4.
10. Galor A, Jabs DA, Leder HA, Kedhar SR: Comparison of antimetabolite drugs as corticosteroid-sparing therapy for noninfectious ocular inflammation. *Ophthalmology*. 2008; 115(10):1826-32
11. Cazabon S, Over K, Butcher J: The successful use of infliximab in resistant relapsing polychondritis and associated scleritis. *Eye (Lond)*. 2005; 19(2): 222-4

Author Information

Asha Ramanathan, MBBS, BSc

Department of Ophthalmology, Royal Glamorgan Hospital

Amit Gaur, MBBS, MD, FRCS

Consultant Ophthalmologist, Royal Glamorgan Hospital