Available online at *www.ujpronline.com*
Universal Journal of Pharmaceutical Research
An International Peer Reviewed Journal
ISSN: 2831-5235 (Print); 2456-8058 (Electronic)

CASE STUDY

POSTERIOR SCLERITIS: REVIEW OF LITERATURE AND FIRST CASE REPORT FROM YEMEN

Essam Yahiya Al-shamahi[1], Sameha A. Al-Eryani[1], Emad Hassan Al-Shamahi[1], Nawal Mohammed Al-Hababi[1], Nashwan Yahya A Al-Shamahi[2], Hassan Abdulwahab Al-Shamahy[2,3]

[1]*Opthalmology Department, Faculty of Medicine and Health Sciences, Sana'a University, Republic of Yemen.*
[2]*Medicine Department, Faculty of Medicine and Health Sciences, Sana'a University, Republic of Yemen.*
[3]*Department of Basic Sciences, Faculty of Dentistry, Sana'a University, Republic of Yemen.*

Article Info:

Article History:
Received: 6 September 2024
Reviewed: 8 November 2024
Accepted: 21 December 2024
Published: 15 January 2025

Cite this article:
Al-shamahi EY, Al-Eryani SA, Al-Shamahi EH, Al-Hababi NM, Al-Shamahi NYA, Al-Shamahy HA. Posterior scleritis: Review of literature and first case report from Yemen. Universal Journal of Pharmaceutical Research 2024; 9(6): 90-95.
http://doi.org/10.22270/ujpr.v9i6.1244

***Address for Correspondence:**
Prof. Hassan A. Al-Shamahy, Department of Basic Sciences, Faculty of Dentistry, Sana'a University, Republic of Yemen. Medical Microbiology and Clinical Immunology Department, Faculty of Medicine and Health Sciences, Sana'a University. Tel: +967-1-239551; E-mail: *shmahe@yemen.net.ye*

Abstract

A rare inflammatory eye illness that involves the posterior regions of the sclera. Posterior scleritis is very rare in children and more common in women. Ocular pain, headaches, and vision loss are some of its frequently vague clinical manifestations. Rheumatic conditions including systemic lupus erythematous (SLE), anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitis and rheumatoid arthritis (RA) are frequently linked to the scleritis pathogenesis. A comprehensive clinical eye exam is necessary to diagnose posterior scleritis because it can mimic many other ocular disorders. Laboratory tests may reveal underlying systemic disorders, such as rheumatic disease and inflammatory markers. A precise diagnosis is aided by imaging, such as magnetic resonance imaging (MRI) and B-scan ultrasonography. Treatment includes non-steroidal anti-inflammatory drugs (NSAIDs), topical corticosteroids for moderate disease, and systemic corticosteroids for severe disease. For refractory instances, biologic therapy has grown in importance. To treat this potentially blinding condition, a multidisciplinary strategy combining rheumatology and ophthalmology is essential. This case report focuses on a 9-year-old boy who has posterior scleritis and no history of rheumatic disorders or other infections or non-infectious diseases.

Keywords: CT scan, posterior scleritis (T-sign), rare episode, Sana'a, ultrasound, Yemen.

INTRODUCTION

A dangerous inflammatory state described scleritis damages the sclera, the white outer layer of the eye. The illness is frequently acquired by co-occurring diseases similar to rheumatoid arthritis or granulomatosis with polyangiitis. Scleritis comes in three diverse forms: diffuse scleritis, the most common type, nodular scleritis, and necrotizing scleritis, the most severe. It's possible that scleritis is the initial sign of connective tissue illness[1]. Also it is rare inflammatory disease of the eye that affects the posterior region of the sclera[2]. With a mean age of beginning in the 40s, posterior scleritis is more frequent in females than in males. It is the rarest type of scleritis, accounting for 3% to 17% of all cases[2-4]. Clinical symptoms are typically monocular and quite vague. Bilateral illness is considerably less common, with binocular cases accounting for about one-third of cases[5]. Ocular pain, which is made worse by eye movements, headaches, and vision loss are typical symptoms[2]. Due to its nonspecific presentation, it is frequently misdiagnosed and may be visually dangerous; diagnosis requires a high index of suspicion and skill[3]. Rheumatic conditions such systemic erythematous lupus (SLE), anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitis, and rheumatoid arthritis (RA) are frequently linked to posterior scleritis[2]. The gold standard test for detecting posterior scleritis is B-scan ultrasonography, which measures scleral thickness with great sensitivity and specificity[6]. Scleral thickness can be determined and disease activity can be tracked with optical coherence tomography (OCT) [Figure 3, and Figure 4][7]. Topical NSAIDs and corticosteroids are typically used as treatment; however, they are often ineffective, necessitating the use of oral NSAIDs. When oral NSAIDs don't work, systemic steroids are necessary, and steroid-sparing medications such mycophenolate or methotrexate can be used for chronically active

conditions or corticosteroid intolerance[7]. Biologic therapy, such as rituximab or tumor necrotic factor-alpha (TNF) inhibitors, may be required in certain situations[7]. Up to one-third of individuals experience recurrence, hence it is advised that ophthalmologists and rheumatologists treat patients together[4,7]. A case of bilateral posterior scleritis that is unrelated to systemic illnesses is shown here.

CASE REPORT

Our Institutional Review Board (IRB) approved the publication of this case report, and the parent patient provided written informed consent for the publication of his son's data. The 1964 Helsinki Declaration and its subsequent amendments were upheld in the study. This case concerns a 9-year-old Yemeni boy who presented with acute visual acuity drop, despite having no clinical symptoms or features suggestive of scleritis. He was afebrile and had stable vital signs at presentation. Examined, he showed no ophthalmoplegia, no pain with extra-ocular eye movement, normal conjunctival and eyelid, and bilaterally equal and light-responsive pupils. During the regular laboratory workup (complete blood count and basic metabolic panel), all inflammatory indicators, including erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), were found to be normal. According to his clinical systemic history, he had no history of parathyroid issues, rheumatic disorders, or any other infections or non-infections illness. His ocular history showed no signs of trauma or clinically significant intraocular inflammation. The refractometry values for his left sphere cylinder and axis were -0.75 and 135 while his right sphere cylinder and axis were -1.0 and 85. With the help of eyeglasses, his visual acuity increased from 5% in both eyes to 20%. Fundus picture revealed a 5 DD gray to white lesion around the optic disc in the left eye (Figure 1). Also, fundus picture revealed a 2 DD gray to white lesion around the optic disc in the right eye (Figure 2).

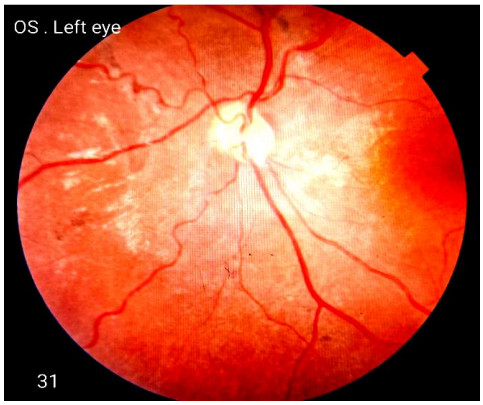


Figure 1: Fundus examination revealed a 5 DD gray to white lesion around the optic disc in the left eye.

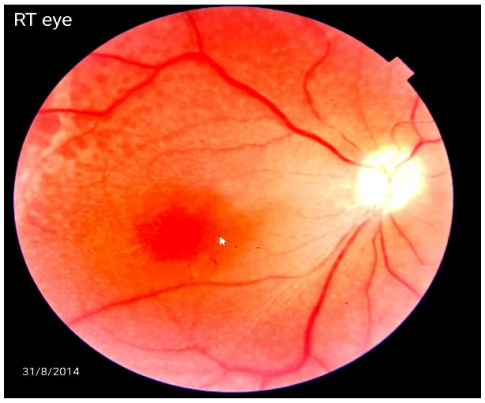


Figure 2: Fundus examination revealed a 2 DD gray to white lesion around the optic disc in the right eye.

Ocular ultrasound was then performed and showed a hyperreflective lesion around the optic disc resembling a choroidal osteoma. CT scan was then performed and revealed no osteoid lesion so choroidal osteoma was ruled out. Re-evaluation by sonagraphy revealed posterior scleritis (T sign). Steroids administered subtenon were used to treat the patient. When compared to previous OCT imaging (Figure 3, Figure 4), his optical coherence tomography (OCT) examination of both eyes, performed eight weeks after treatment of his posterior scleritis flare, revealed stable thickness and no signs of retinal detachment or choroidal edema (Figure 5, and Figure 6).

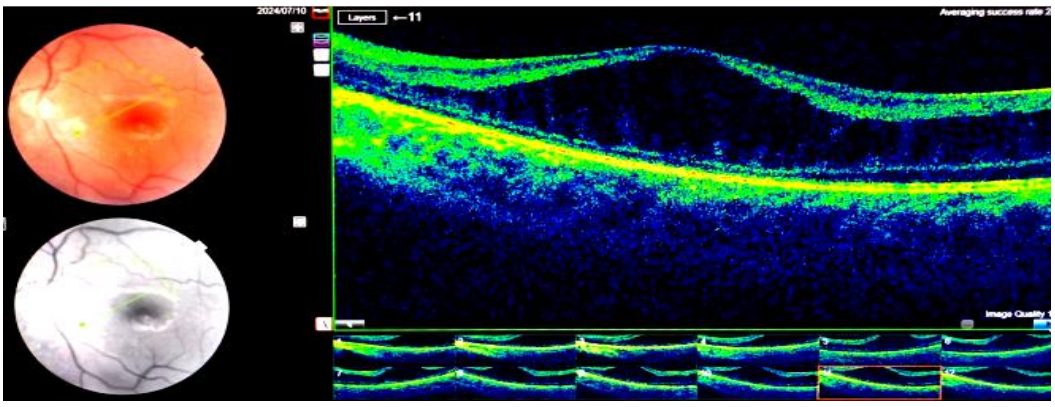


Figure 3: Previous OCT imaging revealed thickness and signs of retinal detachment or choroidal edema for left eye.

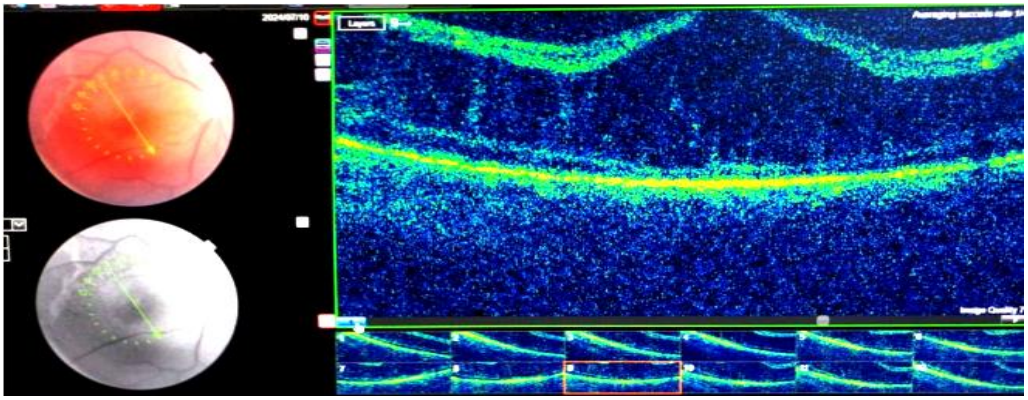


Figure 4: Previous OCT imaging revealed thickness and signs of retinal detachment or choroidal edema for right eye.

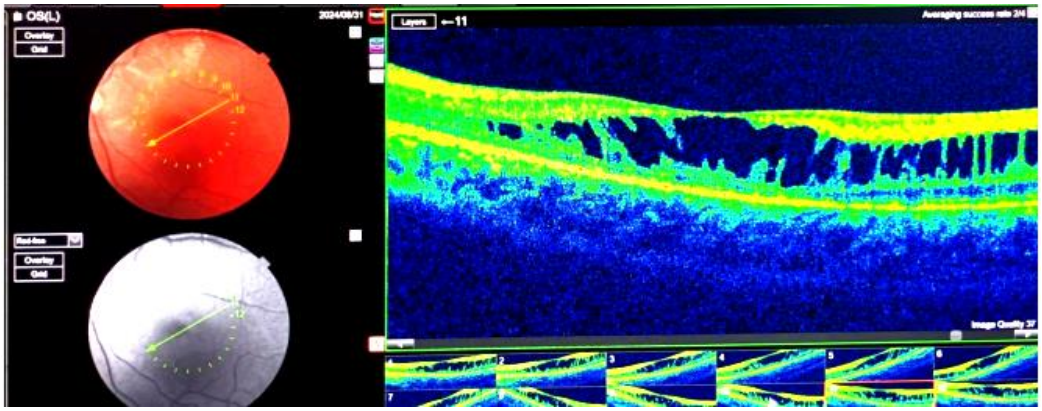


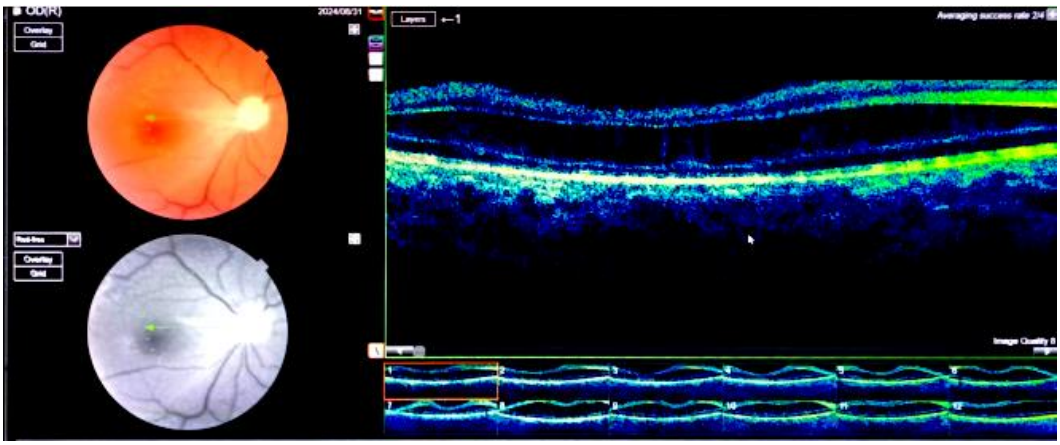
Figure 5: Optical coherence tomography (OCT) examination of both eyes, performed eight weeks after treatment of his posterior scleritis flare, revealed stable thickness and no signs of retinal detachment or choroidal edema for left eye.

DISCUSSION AND REVIEW OF LITERATURE

A rare inflammatory disorder of the eye is called posterior scleritis. It has a wide range of clinical presentations, which can make diagnosis difficult[1]. The male youngster in our study was 9 years old, although other studies have shown that the disease is more frequent in females and that the mean age of appearance is approximately 45 years old[2-4].

The patient in the current study also had bilateral posterior scleritis, which is different from the previous fact that it is typically unilateral but can infrequently impact both eyes in as many as one-third of cases[5].

Eye pain, headaches, ocular movement pain, and, if left untreated, blindness are all signs of posterior scleritis[2]. Conjunctival chemosis, hyperemia, and enlargement of the optic nerve are examples of nonspecific clinical symptoms[6]. If inflammation is exclusively posterior, the eye may also be without apparent redness, which makes identification much more difficult and necessitates a high level of clinical suspicion[3,7]. Our patient had no conjunctival erythema and no bilateral eye pain or ocular movement pain.



Figures 6: Optical coherence tomography (OCT) examination of both eyes, performed eight weeks after treatment of his posterior scleritis flare, revealed stable thickness and no signs of retinal detachment or choroidal edema for right eye.

Even though he was previously diagnosed with posterior scleritis, concurrent anterior scleritis is implausible given his presentation, which lacks obvious eye redness and swelling. In one case study, 59% of patients with posterior scleritis also developed anterior scleritis, indicating that concurrent anterior scleritis is prevalent in posterior scleritis[8]. Because the illness can mimic other ocular pathologies, it is important to have a sharp clinical eye to recognize its unique characteristics. Acute angle closure glaucoma and orbital/preseptal cellulitis are examples of differential diagnosis. Similar presentations can be seen in orbital masses such as rhabdomyosarcoma, leukemia, neuroblastoma, and orbital pseudotumor, commonly referred to as idiopathic orbital inflammation[9,10]. The diagnosis requires a strong index of suspicion[3]. There were no systemic, infectious, or noninfectious events in our patients. According to earlier research, between 40 and 50 percent of cases of scleritis are linked to systemic illnesses, whether they are infectious or not[4,9]. Although SLE and granulomatosis with polyangiitis (GPA) are frequently linked to posterior scleritis, RA is the inflammatory disorder most frequently associated with it[4,11,12]. There is no history of viral infection, trauma, or drug use in our instance. Nonetheless, earlier research indicated that orbital trauma, drugs like bisphosphonates, and infections (specifically, varicella zoster virus (VZV) and herpes zoster virus (HZV)) are among the causes of scleritis[9,13]. In some cases, idiopathic etiology is seen[10].

As hospitalists and ophthalmologists, we might be the first medical professionals that patients with this condition seek out, but frequently, caregivers lack the knowledge and resources necessary to promptly diagnose posterior scleritis. In this patient group, this could lead to incorrect diagnosis, inappropriate therapy, and even blindness. Because patients with autoimmune illnesses are more likely to develop non-infectious posterior scleritis, healthcare providers need to be very careful while treating ocular problems in these patients. Rheumatologists and ophthalmologists typically co-manage these individuals[7]. Imaging studies are essential for determining the underlying pathology and making an accurate diagnosis. Anterior sclera inflammation with conjunctival chemosis, vascular dilatation, and tortuosity may be observed during a slit lamp examination[2]. OCT revealed choroidal thickness in our patients because the disease was active. It is well recognized that OCT can reveal choroidal thickening in cases of active disease and is a helpful tool for monitoring the progression of the condition. When treatment is effective, the thickening will decrease[14]. However, the mainstay of imaging for posterior scleritis is B-scan ultrasonography. It typically exhibits the pathognomonic T-sign, which is fluid in sub-Tenon's space, along with choroidal and scleral thickness larger than 2.5 mm. A scleral thickness of greater than 1.7 mm was suggested by the authors of one retrospective case series, and it was discovered to be more sensitive (87.5%) and specific (88.9%) in identifying posterior scleritis, particularly in cases that were mild and early[6]. It is crucial to remember that, particularly in cases when clinical suspicion is high, a negative T-sign does not rule out posterior scleritis[6].

By providing a more thorough anatomical outline, MRI enhances the diagnostic toolkit and is helpful in situations when ultrasonography is inconclusive[10]. Formalizing the diagnosis and directing future treatment plans can be accomplished by cooperative interpretation of these imaging data[4]. Other laboratory tests, like a serologic test for rheumatic disorders like an anti-nuclear antibody (ANA), rheumatoid factor antibody (RF), anticitrullinated antibody (CCP), and ANCA, can also help diagnose this condition. In addition to acute phase reactants like CRP and ESR, a high RF for RA can also indicate active disease flair. Assessing disease activity can also be facilitated by these indicators[11].

Even though our patients' CBC results were normal, researchers discovered that standard laboratory tests, such as complete blood counts, can show higher white blood cells, and complete metabolic panels can show indications of liver inflammation and renal insufficiency. These results may offer hints regarding underlying systemic conditions such as GPA2, autoimmune hepatitis, and SLE. Therapy depends on a precise diagnosis and a multidisciplinary approach that includes ophthalmology and rheumatology. The therapeutic options for posterior scleritis will be further examined in this case study. First-line treatments include oral non-steroidal anti-inflammatory medications and topical corticosteroids[8,9]. Young patients with unilateral scleritis and those without concomitant systemic illnesses typically respond well to these first-line treatments. Systemic steroids are among the alternative treatments that a significant number of patients with mild-to-moderate illness will require[8,9]. Prednisolone is often started at a dose of 1 mg/kg/day and tapered weekly based on the clinical response, with oral corticosteroids continuing to be the cornerstone of short-term therapy of scleritis[7,8]. Use of pulse IV corticosteroids is essential, particularly when inflammation needs to be controlled quickly and the patient's vision is at risk[8]. Our patient's symptoms improved, but his vision was severely compromised, necessitating the use of IV corticosteroids.

Long-term immunosuppressive treatment is typically necessary for posterior scleritis, which can return frequently[4,9]. Long-term corticosteroids present difficulties because of their numerous adverse effects. In cases of failure or insufficient response to steroids, recurring disease on more than 10 mg/day of oral corticosteroid, and severe side effects of steroids, the use of steroid-sparing medicines is recommended[8]. According to Robertson *et al.*[15], her patient experienced a return of her disease after taking many DMARDs, underscoring the recurrent nature of this condition. According to a study on treatment preferences for scleritis, methotrexate was the most popular first-choice steroid-sparing medication[8]. This was impacted by a number of parameters, including patient tolerance, adjustable dose, and simplicity of weekly administration. On the other hand, it is linked to a quicker relapse[12]. Given their tolerability and quicker steroid-sparing effect than methotrexate,

azathioprine and mycophenolate are additional steroid-sparing drugs to take into account[8]. Another medication is cyclophosphamide, which is mostly utilized in individuals with systemic vasculitis like GPA[7,9]. Its side effect profile, which does not improve efficacy in comparison to other immunosuppressive treatments, is a limiting factor in its utilization[8].

Biologic therapy's focused strategy of modifying the inflammatory cascade has made it extremely helpful in the therapy of posterior scleritis. According to one study, rheumatologists and uveitis specialists' first-choice biologic drug for treating scleritis was TNF-alpha inhibitors[8]. Since they have been demonstrated to be successful in treating treatment-resistant scleritis, TNF-alpha inhibitors like infliximab and adalimumab are excellent choices in this toolbox[8]. On a case-report basis, golimumab has demonstrated efficacy in treating severe refractory posterior scleritis[16]. A further first-line biologic drug that has been demonstrated to be well-tolerated and successful in reducing scleral inflammation in refractory scleritis is rituximab; however, it has a high recurrence rate and necessitates repeated infusions[8,9,16]. In the therapy of scleritis, newer biologic drugs such tocilizumab, an interleukin-6 inhibitor, have showed promise, particularly when used in patients with severe systemic rheumatoid vasculitis who also have several extraarticular disorders[8]. Scleritis can also be effectively treated with janus kinase inhibitors, such as tofacitinib[8]. The immune-mediated inflammatory response to the sclera is disrupted by abatacept, a T-cell costimulator modulator that prevents T-cell activation[17]. Its administration has been demonstrated to significantly induce remission in posterior scleritis, according to data from a case report[18]. Despite having previously had treatment with methotrexate, mycophenolate, rituximab, and infliximab, her patient experienced numerous relapses, according to Robertson *et al.*[15]. The most recent flare-up happened after two weeks of not taking abatacept. The potential effectiveness of biologics in treating refractory cases of posterior scleritis is demonstrated by this example. However, the dearth of solid data from carefully planned and sufficiently powered studies about the use of biologics highlights the necessity of more investigation to develop treatment plans for posterior scleritis and scleritis in general.

Limitation of the study

One of the shortcomings of this study is that it relied on one eye center to study posterior scleritis, and there is a need to conduct a study of cases in all eye centers in Yemen so that we can evaluate this problem in Yemen and build a good understanding of this problem in Yemen as well as its association with auto-immune diseases.

CONCLUSIONS

A rare but dangerous inflammatory disease that affects the sclera, posterior scleritis can seriously impair vision if left untreated. Although less common, bilateral illness has been observed in our patient and may be linked to concurrent anterior scleritis. In order to identify this illness early and stop vision loss, doctors must maintain a high index of suspicion. To customize particular treatment plans, a multidisciplinary approach involving ophthalmology and rheumatology is required. Even though therapy is difficult, the use of biologics becomes a worthwhile consideration when traditional therapies are insufficient, particularly when the disease recurs, as demonstrated in the previous trial. Although biologic therapy has potential for treating posterior scleritis, more extensive research is needed to fully develop evidence-based treatment recommendations.

ACKNOWLEDGEMENT

The work was financed by Sana'a University, Sana'a, Yemen, which the authors would like to thank.

AUTHOR'S CONTRIBUTIONS

Al-shamahi EY: writing the original draft, diagnosing the case. **Al-Eryani SA:** conceptualization. **Al-Shamahi EH:** critical review, diagnosing the case. **Al-Hababi NM:** review and editing, **Al-Shamahi NYA**: Formal analysis, data processing. **Al-Shamahy HA:** data analysis. All authors reviewed and approved the final version of the article.

DATA AVAILABILITY

Upon request, the accompanying author can furnish the empirical data used to bolster the findings of the study.

CONFLICT OF INTEREST

There is no conflict of interest around this work.

REFERENCES

1. John F., Salmon. Episclera and sclera. Kanski's clinical ophthalmology : A systematic approach (9th ed.) 2020. Edinburgh:Elsevier. ISBN 978-0-7020-7713-5.
2. Doumazos S, Kandarakis SA, Petrou P, Karagiannis D, Doumazos L, Georgalas I. Posterior scleritis in a patient with psoriasis masquerading as acute angle closure glaucoma. Case Rep Ophthalmol 2022; 13:717- 23. *https://doi.org/10.1159/000526714*
3. Benson WE: Posterior scleritis. Surv Ophthalmol 1988; 32:297-316. *https://doi.org/10.1016/0039-6257(88)90093-8*
4. Lavric A, Gonzalez-Lopez JJ, Majumder PD, Bansal N, Biswas J, Pavesio C, Agrawal R. Posterior scleritis: Analysis of epidemiology, clinical factors, and risk of recurrence in a cohort of 114 patients. Ocul Immunol Inflamm 2016; 24:6-15. *https://doi.org/10.3109/09273948.2015.1005240*
5. Wen C, Duan H: Bilateral posterior scleritis presenting as acute primary angle closure: A case report. World J Clin Cases 2021; 9:3779-86. *https://doi.org/10.12998/wjcc.v9.i15.3779*
6. Maleki A, Ruggeri M, Colombo A, Asgari S, Look-Why S, Apoorva S, Foster CS. B-scan ultrasonography findings in unilateral posterior scleritis. J Curr Ophthalmol 2022 Apr 16;34(1):93-99. *https://doi.org/10.4103/joco.joco_267_21*
7. Posterior scleritis: A diagnostic challenge 2018. Accessed: February 27, 2024:

https://www.reviewofophthalmology.com/article/posterior-scleritis-a-diagnostic-challenge

8. Dutta Majumder P, Agrawal R, McCluskey P, Biswas J: Current approach for the diagnosis and management of noninfective scleritis. Asia Pac J Ophthalmol (Phila). 2021; 10:212-23. *https://doi.org/10.1097/APO.0000000000000341*
9. Hogan C, Vakros G, Jones R, Bhalla S, McVeigh K: More than meets the eye: A rare case of posterior scleritis masquerading as orbital cellulitis. Cureus 2020; 12:e8177. *https://doi.org/10.7759/cureus.8177*
10. Murthy SI, Sabhapandit S, Balamurugan S, *et al*. Scleritis: differentiating infectious from non-infectious entities. Indian J Ophthalmol 2020; 68:1818-28. *https://doi.org/10.4103/ijo.IJO_2032_20*
11. Roberts F. The sclera. Eye Pathology. Heegaard S (ed): Springer, Berlin, Germany 2014; 155-72. *https://doi.org/10.1007/978-3-662-43382-9_4*
12. Promelle V, Goeb V, Gueudry J. Rheumatoid arthritis associated episcleritis and scleritis: An update on treatment perspectives. J Clin Med 2021; 10:2118. *https://doi.org/10.3390/jcm10102118*
13. Fraunfelder FW, Fraunfelder FT: Adverse ocular drug reactions recently identified by the National Registry of Drug-Induced Ocular Side Effects. Ophthalmol 2004 111:1275-9. *https://doi.org/10.1016/j.ophtha.2003.12.052*
14. Vermeirsch S, Testi I, Pavesio C: Choroidal involvement in non-infectious posterior scleritis. J Ophthalmic Inflamm Infect 2021; 11:41. *https://doi.org/10.1186/s12348-021-00269-9*
15. Robertson R, Ansari F A, Gafoor S, *et al*. Posterior Scleritis: A case report and literature review of the management of the condition. Cureus 16(6): e61537. *https://doi.org/10.7759/cureus.61537*
16. Sota J, Girolamo MM, Frediani B, Tosi GM, Cantarini L, Fabiani C: Biologic therapies and small molecules for the management of non-infectious scleritis: A narrative review. Ophthalmol Ther 2021, 10:777-813. *https://doi.org/10.1007/s40123-021-00393-8*
17. Dubois EA, Cohen AF: Abatacept. Br J Clin Pharmacol. 2009; 68:480-1. *https://doi.org/10.1111/j.13652125.2009.03502.x*
18. Comacchio F, Cecchin V, Martini G, Meneghel A, Zannin ME: Posterior scleritis in a teenager responding to abatacept. EC Ophthalmol 2018; 4:384-7.