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DIAGNOSTIC AND THERAPEUTIC CHALLENGES OF POSTERIOR SCLERITIS MIMICKING CHOROIDAL MELANOMA

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Abstract

Introduction: *Posterior scleritis is a rare form of scleritis. Posterior scleritis occurs in 2-12% of cases of scleritis, 64,4-71,1% occur in females. Its clinical features may be confused with choroidal pathologies like choroidal melanoma. Posterior scleritis is 1,5% from 400 lesions simulating choroidal melanoma. This case report presents a diagnosis and therapeutic challenges of posterior scleritis mimicking choroidal melanoma. Diagnosis is made based on history taking, complete ophthalmology examination, and ancillary test.*

Case Report: *A-49-year-old woman came to outpatient clinic with gradual vision loss and ocular pain. She has history of hypothyroid disease. Her visual acuity (VA) was 6/15 in the right eye and 6/9,5 in the left eye. Posterior segment examination revealed disc edema. B-Scan Ultrasonography showed choroidal thickening without T-sign. She was diagnosed with posterior scleritis and received systemic corticosteroids. At the 4-month follow-up, her VA was 6/60 in the left eye, and exudative retinal detachment was found on funduscopy, B-scan ultrasonography showed a hyperechoic subretinal mass with overlying subtenon fluid reflection. In a month, B-Scan Ultrasonography of the left eye showed a larger nodular mass with characteristic 'T-Sign', OCT macula showed subretinal fluid and MRI showed thickening of the left posterior sclera. Posterior scleritis was identified, and then the patient received oral methotrexate and high-dose corticosteroid. At the 2-week follow-up, VA of the left eye improved to 6/18 and exudative retinal detachment has resolved. B-Scan ultrasonography showed scleral thickness was decreased but T-sign still exists.*

Discussion: *Posterior scleritis can mimic choroidal melanoma, the diagnosis of posterior scleritis is based on clinical presentation, eye examination, and ancillary testing. B-scan ultrasound is useful for confirming posterior scleritis. Posterior scleritis can be treated with steroids or a combination of steroids and antimetabolite if steroid treatment fails or inflammation recurs after tapering the steroids.*

Conclusion: *Posterior scleritis has variable clinical presentation and it may mimic choroidal melanoma. High-suspicion clinical assessment combined with ancillary testing is essential for diagnosis. A combination of steroid and antimetabolite therapies is useful in cases with inadequate response to steroids alone.*

Keywords: Scleritis, Posterior Scleritis, Choroidal Melanoma

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INTRODUCTION

Scleritis is inflammation of the sclera, scleritis can be classified by location (anterior and posterior), severity (necrotizing and non-necrotizing), and type of inflammation of the sclera (diffuse and nodular). Posterior scleritis is a vision threatening that occurs in the posterior ora serrata. The incidence rate of scleritis is estimated at 3.4–4.1 per 100,000 people, with a prevalence of 5.2 per 100,000 residents in America.[1–4]

In posterior scleritis, which is present in 60% of cases, anterior inflammation without anterior scleritis is uncommon. Decreased visual acuity, boring pain, and tenderness to palpation were common symptoms mentioned, but only 30% to 40% of patients complained of pain, whether it was mild or absent. Choroidal detachment and folds, subretinal fluid, and optic disc edema are examples of possible symptoms. Many times, posterior scleritis is misdiagnosed as choroidal melanoma due to its unusual signs and symptoms.[1–4]

In a B-Scan ultrasound for scleritis, the T-sign is typically present, whereas it is occasionally absent in atypical posterior scleritis. Intraocular tumors can be eliminated by magnetic resonance imaging (MRI) and optical coherence tomography (OCT). Sixty percent of posterior scleritis cases are idiopathic, and 40% are associated with systemic or autoimmune disease. The most common systemic or autoimmune-associated disease is rheumatoid arthritis followed by systemic lupus erythematosus. The incidence of posterior scleritis in hypothyroidism has not been reported up to this point. Management attempts to reduce or suppress immune reactions. Steroids are effective to treat posterior scleritis; however, immunomodulators can also be used when the condition of the inflammatory sclera worsens or does not improve.[2,3,5–7]

This case report discusses the diagnosis and therapeutic challenges of posterior scleritis that mimics choroidal melanoma.

CASE REPORT

A 49-year-old woman came to an outpatient clinic with gradual vision loss in both eyes six months ago, accompanied by red eyes and ocular pain. She has history of hypothyroidism on treatment with levothyroxine. On the first examination, the visual acuity in her right eye was 6/15; with correction S-0,50, it became 6/9 and 6/9.5 on her left eye. The Anterior segment of both eyes was normal and no vitreous cells were found. Scleras were hyperpigmented with no scleral thinning, the optic nerve head was edema, and on B-Scan ultrasound, there was choroidal thickening with minimal T-sign on right eyes and without T-Sign on left eye. A laboratory workup was performed, and the results were ANA Test (0,50 ratio), Free T4 was within normal limits (1,24 ng/dL), and RA factor was negative, whereas CRP (12.09 mg/dL) and TSH (14,92 µU/mL) were increased. She was diagnosed with bilateral posterior scleritis with hypothyroidism and treated with steroid 40mg which tapered slowly by 5-10 mg/week and Levothyroxine 50mg.

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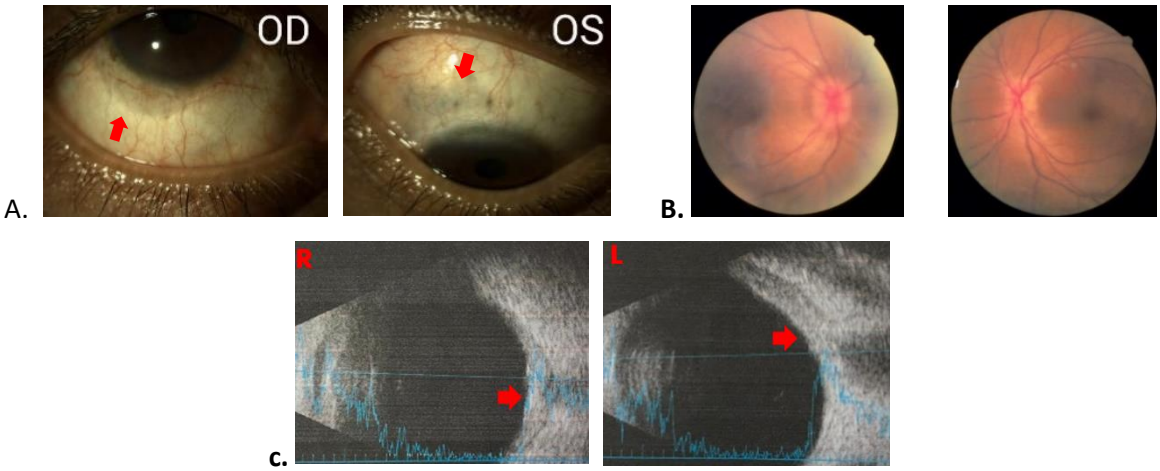

Figure 1. The Clinical condition at the first examination. A. Anterior segment photograph reveals scleral hyperpigmentation without scleral thinning on both eyes. (red arrow). B. Fundus photograph showed optic nerve head edema of both eyes. C. B-scan ultrasound showed choroidal hyperechoic with minimal T-sign on the right eye and without T-sign on the left eye (red arrow)

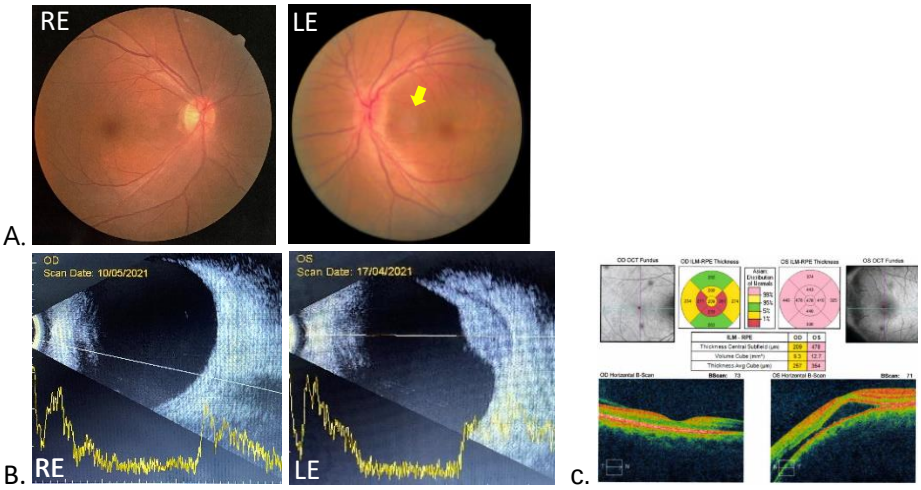

Figure 2. The Clinical condition at 4-month follow-up. A. Fundus photography showed normal right eye and optic nerve head edema with hyperpigmentation which extends to the macula on the left eye (arrow). B. B-Scan Ultrasound showed hyperechoic subretinal mass on the left eye with unclear T-sign. C. Macula OCT showed subretinal fluid on the left eye.

The patient complained of severe visual impairment and clinical examination found that the visual acuity was hand movement. No vitreous cells were found. There was left eye exudative retinal detachment. The choroidal nodular mass in the left eye was larger on the B-Scan ultrasonography, and there was fluid accumulation in subtenon space (a "T-sign"). Scleral thickening and subtenon fluid were detected in the left eye during an orbital MRI with contrast, which was considered to be a sign of posterior scleritis. Based on the results of funduscopy examination and MRI, she was diagnosed with LE exudative retinal detachment and optic nerve head edema due to posterior scleritis. The oral prednisone dose was increased to 60 mg (1 mg/kg/day) tapered 10mg/week and methotrexate 15 mg/week was added.

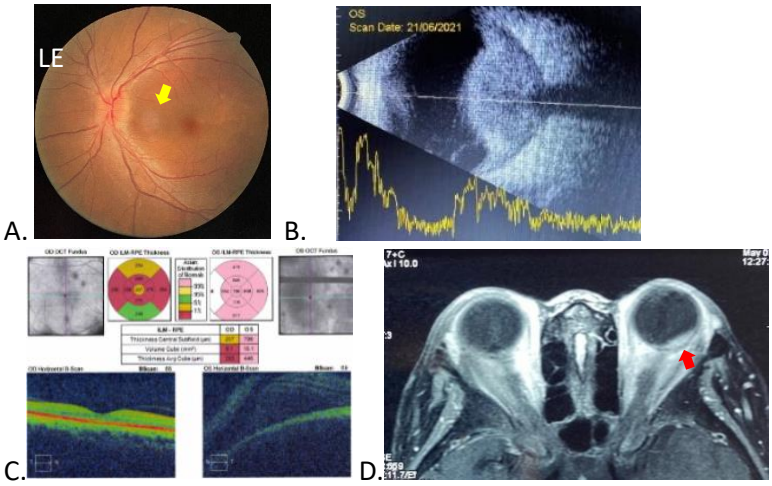

Figure 3. The Clinical condition of 5-month follow-up of left eye. A. Fundus photograph showed optic nerve head edema with hyperpigmentation which extends to macula (arrow) B.B-Scan Ultrasound showed scleral thickening and fluid accumulation in sub-tenon space. C. Macular OCT showed increased subretinal fluid in the left eye. D. Orbital MRI showed scleral thickening with T-sign (arrow)

At 1-month follow-up, the left vision became 6/18, hyperpigmentation and optic nerve head edema persisted, but the exudative retinal detachment was resolved. T-sign on B-Scan Ultrasound was still present despite decrease in scleral thickness. Papilledema was still visible on the optic nerve head OCT, however macular OCT showed resolved subretinal fluid.

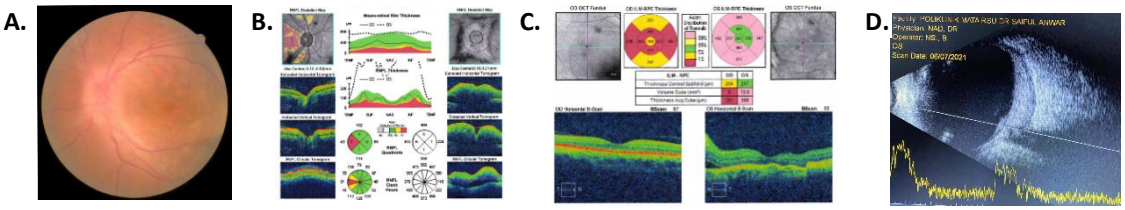

Figure 4. The Clinical condition of 6-month follow-up. A. Fundus Photograph showed hyperpigmentation lesions still exist. B. Optic nerve head OCT showed Optic nerve head edema still exist on the left eye. C. Macular OCT showed subretinal fluid has been resolved. D. B-Scan Ultrasound showed decreased scleral thickness and T-sign.

DISCUSSION

Most posterior scleritis causes are idiopathic. Inflammation and systemic autoimmune disorders can also contribute to posterior scleritis. Rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) are the two most prevalent autoimmune disorders that result in posterior scleritis. Sarcoidosis, polyarteritis, thyroid disease, Vogt-Koyanagi-Harada, and other systemic disorders, in addition to RA and SLE, can result in posterior scleritis.[2,3] A history of hypothyroidism was discovered in the patient, which may be the cause of posterior scleritis. Xu et al. , in their research, among 77 scleritis patients, 8 scleritis patients (10,4%) had a diagnosis of thyroid disease. [8]

The anterior region of both eyes had a hyperpigmented lesion that seemed to be scleromalacia. Scleral melanocytosis is a hyperpigmentation disorder found in the sclera. These two conditions can be distinguished

by the presence of scleral thinning. Posterior scleritis is an inflammation of the sclera, posterior to the ora serata, that may be sight-threatening and may cause severe pain and tenderness to palpation. Clinically, posterior scleritis can manifest as exudative retinal detachment, optic nerve head edema, macular edema, and choroidal folds, which are the most frequent abnormalities. Choroidal melanoma has a dome-shaped nodule or mass under the retinal pigment epithelium, so that it appears as a hyperpigmented mass. In this case, the patient complained of blurry vision accompanied by red eye and ocular pain. On examination, we found scleral hyperpigmentation and optic nerve head edema in both eyes. On optic nerve head and macular OCT examinations showed optic nerve head edema. After treatment with steroids, a dome-shaped mass with hyperpigmentation on the optic nerve head that extends to the macula was found in the left eye. That sign of posterior scleritis can mimic choroidal melanoma. Choroidal melanoma typically starts with hyperpigmentation dome-shaped lesions and may cause serous detachment, this manifestation is also found in posterior scleritis. Ancillary testing such as ultrasonography and CT or MRI scan evaluation helped to differentiate. [9–13]

The diagnosis of posterior scleritis is based on clinical presentation and examinations. A B-Scan ultrasound examination is needed to determine the involvement of the posterior sclera, which shows thickening of the sclera and accumulation of fluid in the sub-Tenon space, which appears as a T-sign. Ultrasound helps to assess the involvement of adjacent structures, including the choroid, retina, ciliary body, extraocular muscles, and orbit. Patients with choroidal melanoma show a collar stud shape or a mushroom shape with low to medium spikes. Posterior scleritis can mimic choroidal melanoma; in this case, a B-Scan ultrasound examination showed scleral thickening that looked like a mass in the choroid, while an ultrasound two months later showed an accumulation of fluid in the subtenon space. Laboratory tests such as C-reactive protein (CRP), antinuclear antibody (ANA), rheumatoid factor, and antineutrophil cytoplasmic antibodies (ANCA) are needed to find causes of posterior scleritis. Complete blood counts, blood sugar levels, liver function, and kidney function are tested to monitor the side effects of steroids and methotrexate. An orbital CT or MRI examination can be done to eliminate choroidal melanoma. MRI in choroidal melanoma appears as a solid mass hyperintense nodule, whereas in posterior scleritis shows scleral thickening and sub-Tenon fluid as seen in the 65755 patients. [2,3,12–15]

Posterior scleritis treatment aims to reduce the inflammatory response. Systemic corticosteroids are used to treat posterior scleritis; they started at 1 mg/kg/day and gradually tapered off. Immunomodulators can be administered if the condition worsens or fails with corticosteroids. Antimetabolite groups such as methotrexate, mycophenolate mofetil, or azathioprine are used. Long-term treatment is required to achieve remission and manage the underlying systemic condition. In this case, there was no clinical improvement with corticosteroids; she has decreased left eye visual acuity accompanied by optic nerve head edema, subretinal fluid accumulation, and exudative retinal detachment. After the patient was given corticosteroids and methotrexate, her visual acuity improved and her optic nerve head edema decreased. On B-Scan, ultrasound showed reduced scleral thickness compared to previous scans, but still found a T-sign and reduced subretinal fluid, which was seen on macular OCT. Diagnosing posterior scleritis alone by ophthalmological examination is challenging. Ultrasonography and other supporting exams are necessary.

CONCLUSION

The diagnosis of posterior scleritis requires history taking, eye examination, and an ancillary examination. Decreased visual acuity and a hyperechoic subretinal mass without a T-sign on ultrasound are signs and symptoms of posterior scleritis that were found in this patient which is similar to choroidal melanoma. The combination of steroids and immunomodulators may be used to treat posterior scleritis that does not respond to steroid therapy alone. Collaboration with other departments such as internal medicine is important to treat the patient with posterior scleritis. In this patient, there was improvement in vision, decreased scleral thickness, and reduced macular edema after being treated with steroids and methotrexate.

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