Case report

Eyes as Windows: Ocular Manifestations Paving the Path to Sarcoidosis Diagnosis

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Abstract

A 32-year-old male presented with sudden, painless vision loss in both eyes. Examination revealed impaired visual acuity, vitreous inflammation, and macular edema. Systemic investigations showed elevated inflammatory markers and angiotensin-converting enzyme (ACE) levels, suggesting granulomatous disease. Tuberculosis was excluded, and a diagnosis of ocular sarcoidosis was established. Treatment with corticosteroids led to significant improvement in visual acuity. Ocular sarcoidosis is a common manifestation of systemic sarcoidosis, a multisystem disease characterized by noncaseating granulomas. Ocular involvement occurs in 12-76% of systemic sarcoidosis cases, with uveitis and conjunctival nodules being common findings. High suspicion and adherence to IWOS criteria are crucial for diagnosis.

Keywords: Ocular Sarcoidosis, Uveitis, Mutton fat KPs, Auto-immune disease

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Received 17 February 2025; Accepted 21 February 2025

1. Introduction

Ocular sarcoidosis is a significant manifestation of sarcoidosis, a systemic granulomatous disease characterized by the formation of non-caseating granulomas. This condition affects approximately 25% of patients with sarcoidosis, leading to potentially severe ocular complications that can significantly impair vision and quality of life. The most common form of ocular involvement is uveitis, particularly posterior uveitis and panuveitis, which can present with symptoms such as blurred vision, eye pain, photophobia, and redness[1]. Understanding the etiology, diagnosis, treatment options, and prognosis of ocular sarcoidosis is essential for effective management. The clinical presentation of ocular sarcoidosis can be diverse. Uveitis, an inflammation of the uveal tract, is the most frequently observed condition, manifesting in various forms, including anterior, intermediate, and posterior uveitis. Symptoms may vary from mild irritation to severe pain and vision loss[2]. In addition to uveitis, patients may experience other ocular complications, such as conjunctival granulomas, retinal involvement, and optic nerve damage. Complications like glaucoma, cataracts, and retinal detachment can occur, particularly in cases where inflammation is inadequately controlled. Diagnosing ocular sarcoidosis involves a comprehensive clinical assessment, typically conducted by an ophthalmologist[3]. The International Workshop on Ocular Sarcoidosis (IWOS) has established diagnostic criteria that emphasize the necessity of uveitis, compatible systemic findings, and the exclusion of other potential causes of ocular inflammation[4]. A detailed patient history is crucial, along with a thorough physical examination. Systemic symptoms, including respiratory issues, skin lesions, or lymphadenopathy, often accompany ocular manifestations and can guide the diagnostic process.

Imaging techniques, such as chest X-rays or high-resolution computed tomography (CT) scans, are often utilized to assess for pulmonary involvement, a common finding in sarcoidosis[5]. Laboratory tests may include serum angiotensin-converting enzyme (ACE) levels, which can be elevated in sarcoidosis, although this marker is not definitive[6]. Ocular sarcoidosis treatment involves corticosteroids, which can be administered topically, systemically, or via intravitreal injections. Immununosuppressive agents like methotrexate or biologics may be considered for non-responsive or severe cases, under medical supervision[3, 8]. Regular follow-up is essential to monitor for complications and assess the effectiveness of treatment.

In addition to pharmacological interventions, supportive care, including patient education and monitoring for complications, plays a critical role in managing ocular sarcoidosis. Multidisciplinary

collaboration between ophthalmologists, pulmonologists, and other specialists is often necessary to provide comprehensive care[9].

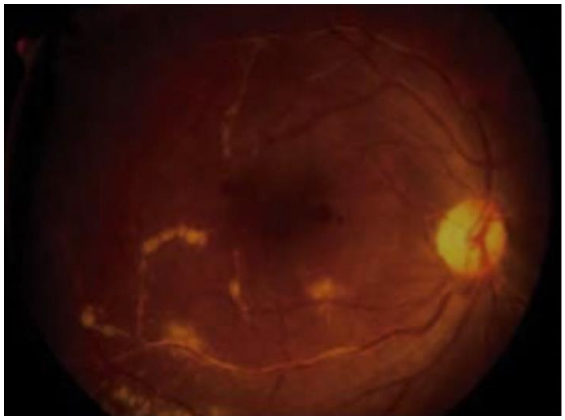
The prognosis for ocular sarcoidosis varies widely. Some patients may experience a self-limiting course, while others may face recurrent episodes that require ongoing management[10]. Long-term follow-up is crucial to monitor for chronic complications and to tailor treatment strategies based on individual patient's responses. Studies have indicated that early intervention can significantly improve visual outcomes and overall quality of life for patients with ocular sarcoidosis[11].

Ocular sarcoidosis requires a multidisciplinary approach for effective management. Early diagnosis and timely intervention are crucial to preserving vision and quality of life. Adhering to International Workshop on Ocular Sacroidosis, (IWOS) guidelines can refine treatment protocols and improve patient outcomes significantly.

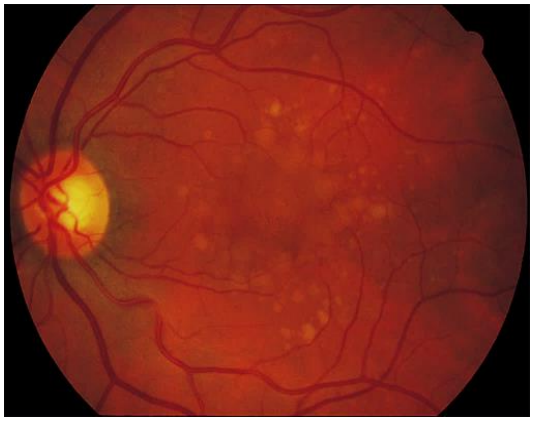
2. Case Presentation

A 32-year-old married male, resident of Rawalpindi, presented to the outpatient department (OPD) in January 2023. The patient reported a sudden onset of painless decreased vision in both eyes that had developed one week prior to his visit. Notably, there was no associated ocular pain, floaters, or flashes of light. His past medical and surgical histories were unremarkable, and he had no significant past ocular history. The family history did not reveal any hereditary eye conditions. The patient is a non-smoker and does not use any addictive substances, identifying as part of a middle-income group.

On Examination, Visual acuity was counting fingers at 3 meters in right eye and 6/36 in left eye. External examination showed normal eyelids without edema or lesions, and the conjunctiva appeared clear with no signs of inflammation. The sclera was also normal, and the cornea was clear without opacities. Intraocular pressure was measured at 15 mmHg in both eyes. The anterior segment examination demonstrated an unremarkable iris with no granulomas, round and reactive pupils, and normally formed anterior chamber. AC was clear without cells or flare. In right eye, vitreous revealed vitritis, vitreous cell +3, vitreous strings & Snow balls in inferior vitreous and no such finding in left eye. Fundoscopic examination revealed Mild Macular Edema, Sub-retinal macular solitary infiltrate temporal to fovea, Peripheral vasculitis, perivenous sheathing, candle wax drippings along inferior vascular arcade, a normal optic nerve head, while fundus examination of left eye showed Mild subretinal infiltrates with macular edema. Special tests, including fluorescein angiography and optical coherence tomography (OCT), were conducted, revealing Macular edema along with vitreous inflammation in right eye and mild macular edema in left eye.



(a)



(b)

Figure 1: Fundus photographs of both eyes of patient (a, b).

Following a comprehensive evaluation by a multidisciplinary team of medical specialists, the diagnosis of ocular sarcoidosis was further substantiated. On physical examination, he appeared to be of average height and build, with normal vital signs: blood pressure was 120/80 mmHg, pulse rate was 80 beats per minute, respiratory rate was 18 breaths per

minute, temperature was 98.6°F, and oxygen saturation was 96% on room air. There were no signs of pallor, koilonychia, or clubbing. Thyroid and lymph nodes were not palpable. The systemic investigations demonstrated a normal complete blood count (CBC), indicating no significant hematological issues. However, the erythrocyte sedimentation rate (ESR) was elevated at 39 mm, and C-reactive protein (CRP) levels were also raised at 25 mg/L, suggesting an ongoing inflammatory process. Renal function tests (RFT) were within normal limits, indicating no renal impairment. Notably, the angiotensin-converting enzyme (ACE) level was elevated at 94 U/L (normal range: 9-67 U/L), Both prothrombin time (PT) and activated partial thromboplastin time (APTT) were normal, ruling out significant coagulation issues. The Quantiferon TB Gold test was negative, with a result of 0.01 IU/ml, effectively excluding active tuberculosis. Serum calcium levels were normal at 2.61 mmol/L. Imaging studies, including high-resolution computed tomography (HRCT) and X-ray of the sacroiliac (SI) joints, returned normal findings. These results collectively support the suspicion of ocular sarcoidosis while excluding other potential causes. Three ocular signs and two positive laboratory test, confirmed the diagnosis of ocular sarcoidosis was according to IWOS criteria. The patient was treated with a posterior sub-Tenon injection of triamcinolone acetonide in the right eye, administering 1 ml of a 40 mg/ml solution to reduce inflammation. Additionally, oral prednisolone was prescribed at a dosage of 1 mg/kg in divided doses for three weeks, followed by a gradual tapering over the subsequent month along with proton pump inhibitors for gastric protection. The patient is on close follow-up and has shown significant improvement. Currently, he is asymptomatic, with bilateral visual acuity measured at 6/6, indicating excellent recovery and stabilization of his ocular condition.

3. Discussion

Ocular sarcoidosis is a significant manifestation of systemic sarcoidosis, affecting approximately 25% of the patients. This case highlights a typical presentation in a 32-year-old male who experienced sudden, painless vision loss, a common symptom associated with this condition. The elevated inflammatory markers, specifically the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), along with an increased angiotensin-converting enzyme (ACE) level, are indicative of the underlying inflammatory process characteristic of sarcoidosis[12, 13].

The negative result from the Quantiferon TB Gold test was crucial in excluding active tuberculosis, a common differential diagnosis given the overlap in ocular manifestations. This differentiation is vital, as both conditions can present with similar symptoms, including uveitis and vision changes[14]. Normal serum calcium levels further exclude hypercalcemia, which can be associated with sarcoidosis but was not evident in this patient[15]. Imaging studies, such as high-resolution computed tomography (HRCT) and X-ray of the sacroiliac joints, did not reveal any significant abnormalities, indicating that ocular symptoms were the primary concern at the time of presentation[16].

The management of ocular sarcoidosis typically involves corticosteroids to control inflammation. In this case, a posterior sub-Tenon injection of triamcinolone acetonide was administered, which is a well-accepted approach for localized inflammation. This method allows for targeted delivery of steroids to the affected area, minimizing systemic side effects[17]. Additionally, the patient was placed on oral prednisolone at a tapering dose, following standard treatment protocols for managing sarcoidosis[18]. The introduction of omeprazole to protect against gastrointestinal side effects from prolonged corticosteroid use demonstrates a comprehensive approach to care, addressing potential complications associated with steroid therapy[19]. The patient's follow-up results have been encouraging, with a current bilateral visual acuity of 6/6 and an asymptomatic status. This positive outcome underscores the effectiveness of the treatment regimen in managing ocular sarcoidosis and preventing complications such as vision loss[20, 21, 22].

4. Conclusion

In conclusion, this case illustrates the importance of early diagnosis and appropriate management strategies for ocular sarcoidosis. Continuous monitoring and follow-up are essential to ensure the long-term stability of the patient's condition and to address any potential recurrences of symptoms. Future research should focus on optimizing treatment protocols and exploring long-term outcomes in patients with ocular sarcoidosis to enhance management strategies further.

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