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Case Report *Neuroophthalmology*

Sarcoidosis masquerading non-arteritic ischemic optic neuropathy

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ABSTRACT

Sarcoidosis, a multisystem granulomatous disease, can occasionally involve the optic nerve, often presenting as optic neuritis. However, its association with Non-Arteritic Anterior Ischemic Optic Neuropathy (NAAION) is rare and can create diagnostic challenges. Here, we report a case of a 52-year-old male who initially presented with sudden visual loss in his left eye, diagnosed clinically as NAAION with a disc at risk in the right eye (RE). He was managed with antiplatelets and steroids. Three months later, he experienced vision loss in the RE with evidence of bilateral intraocular inflammation. Imaging, including Fundus Fluorescein Angiography and Indocyanine Green Angiography, revealed choroidal granulomas, raising differential diagnoses of Sarcoidosis, Vogt-Koyanagi-Harada disease, and Tuberculosis. Intravenous methylprednisolone followed by oral steroids resulted in visual improvement in both eyes. This case underscores the importance of comprehensive assessment and imaging to identify atypical presentations of sarcoidosis masquerading as NAAION, ensuring timely intervention and improving visual outcomes.

Keywords: Disc edema, Fundus fluorescein angiography, Non-arteritic ischemic optic neuropathy, Optic neuritis, Sarcoidosis

INTRODUCTION

Sarcoidosis is a granulomatous multisystem disease of unspecified etiology, and the most common complication in the eye is uveitis. However, involvement of the optic nerve has been documented in 1-2% of subjects of sarcoidosis.[1] Association of non-arteritic anterior ischemic optic neuropathy (NAAION) with granulomatous diseases can pose a diagnostic dilemma as well as a treatment challenge. Therefore, detailed clinical evaluation and appropriate investigation can refine our diagnosis. This case report highlights the important clinical clues and course of management strategy in a case of sarcoidosis masquerading as NAAION.

CASE REPORT

A 52 year old man presented with sudden blurring of vision in left eye (LE) since one day. His best corrected visual acuity (BCVA) in the right eye (RE) was 20/20 and LE was 20/60. He had impaired color vision and relative afferent pupillary defect grade 2. He had a history of chronic alcoholism with no other systemic manifestations. His anterior segment

findings were within normal limits in both eyes. Fundus examination of RE revealed a small cupless hyperemic disc and LE pallid disc edema, with the rest of the background normal [Figure 1]. Humphrey visual field examination showed RE within normal limit and LE with advanced field defect [Figure 2]. Optical coherence tomography angiography gave a picture of deficient perfusion in LE [Figure 3]. Considering his age and with no systemic illness, magnetic resonance imaging brain with orbit with contrast was advised, which showed no evidence of any abnormal lesion [Figure 4]. Depending on clinical evaluation, the patient was diagnosed as LE NAAION with RE disc at risk, and the patient was started on antiplatelets and oral steroids.

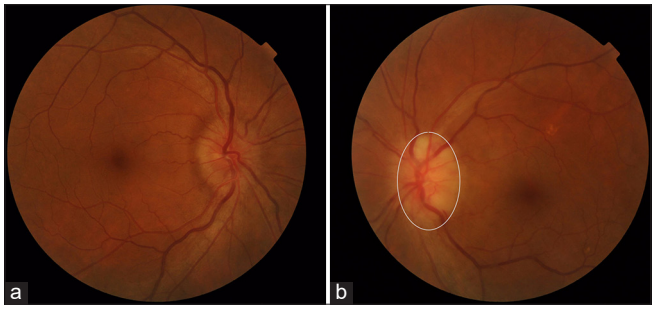

Figure 1: (a) Right eye revealed small cup-less hyperemic disc, (b) Left eye shows pallid disc edema (yellow circle) with rest of background normal.

After three months, his RE developed blurring of vision with BCVA RE 20/250 and LE 20/60. There was a history of typhoid fever one month back and a 7 kg weight loss in the past two months. He also had a history of seizures with unconsciousness three weeks back. On anterior segment examination, both eyes showed the presence of diffuse non-granulomatous keratic precipitates (KPs) and vitreous cells 2+ [Figure 5]. Posterior segment evaluation gave a picture of RE disc edema with a suspected yellowish lesion in the peripapillary region, with few choroiditis lesions noted in midperiphery inferiorly and disc pallor in LE.

Fluorescein angiography showed multiple focal areas of pinpoint hyperfluorescence as well as multiple small choroidal granulomas at the posterior pole came into the picture with indocyanine green angiography [Figure 6]. High-resolution computed tomography chest showed fibrous bands. The complete hemogram and qauntiferion gold were normal. Serum angiotensin converting enzyme was normal; however, there was evidence of dry eye in Schirmer's test 1.

Differential diagnoses of sarcoidosis, Vogt-Koyanagi-Harada disease, and tuberculosis were considered in both eyes, with a previous history of NAAION in LE.

The patient was initiated on corticosteroid pulse therapy (intravenous methylprednisolone 1 g, on three consecutive days) and followed by oral corticosteroids (prednisolone 1 mg/kg body weight) for 11 days. The patient reported

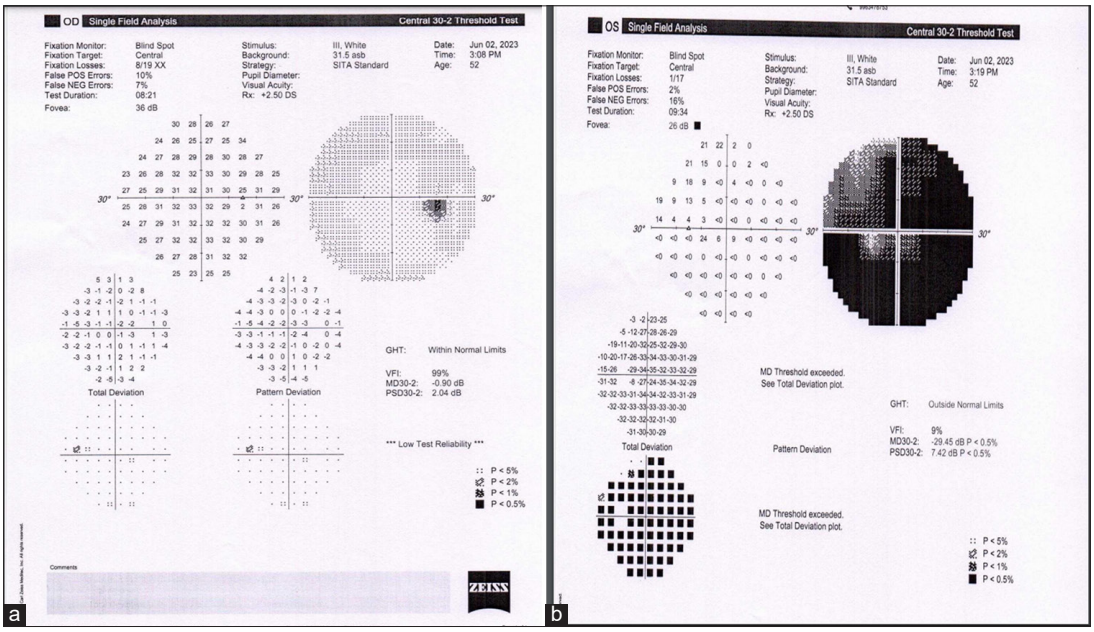

Figure 2: (a) Humphrey visual field examination showed right eye (OD) within normal limits, (b) Left eye (OS) shows advanced field defect.

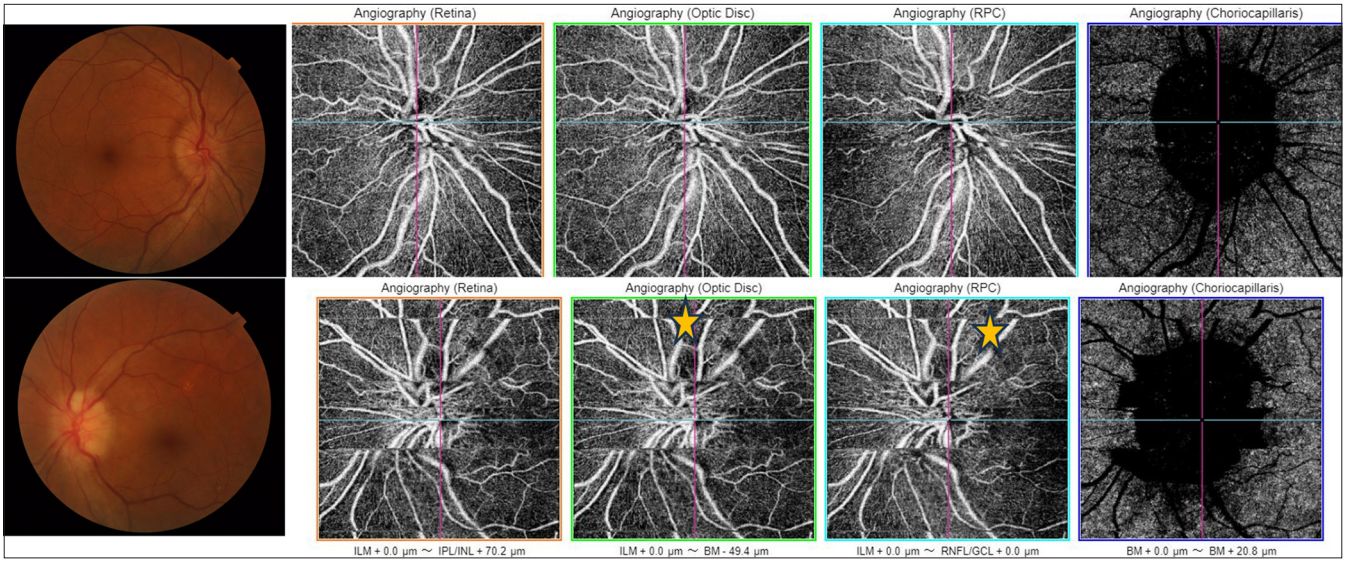

Figure 3: Optical coherence tomography angiography gave a picture of deficient areas of perfusion in left eye as marked by yellow stars. RPC: Retinal pigment cell, ILM: Internal limiting membrane, IPL: Inner plexiform layer, BM: Basement membrane, RNFL: Retinal nerve fibre layer, GCL: Ganglion cell layer.

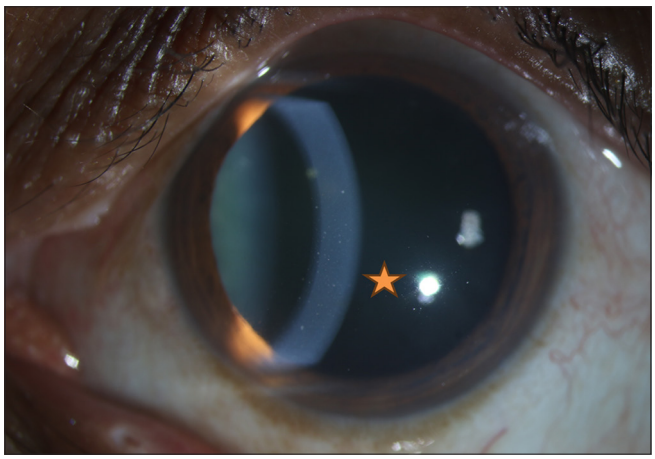

Figure 4: Anterior segment examination of both eyes showed the presence of diffuse non granulomatous keratic precipitates, as shown by yellow star, and vitreous cells 2+.

improvement and with reduced anterior chamber reaction and disc edema in RE. At the end of 2 weeks, his BCVA improved to 20/160 in RE and 20/60 in LE. The patient was shifted to immunosuppressants along with a tapering dose of oral steroids, and after one month, his BCVA in the RE improved to 20/30, but LE vision was stable at 20/60. The patient is kept on long-term low-dose immunomodulators.

DISCUSSION

Sarcoidosis is an auto-inflammatory disorder due to unspecified etiology identified by the enhanced immune reaction, which causes tissue infiltration with inflammatory cells, resulting in the formation of granuloma and subsequent fibrosis.[2] It is common in females, mostly young adults, but may affect any age group.[3] Ophthalmic complications arise in some 10-50%,[4] the most common being anterior uveitis with mutton fat KPs and Koeppe iris nodules, and vitritis leading to the string of pearls.[3]

Nervous system affection is atypical and accounts for 5-10% of published retrospective series.[5] Sarcoid-related optic neuropathy commonly presents like an optic neuritis picture. Optic nerve affection resulting from compressive lesions or from direct granulomatous infiltration is rare.[6]

However, in the present literature, association of NAAION with granulomatous infiltration of the optic nerve is not yet documented, and hence, the pathophysiology is not well understood.

Kidd *et al.*[7] in his large series of patients with sarcoidosis-associated optic neuropathy has shown that half of the patients were not having clinical characteristics of intraocular inflammation, and imaging features were not contributory.

However, in another study by Koczman *et al.*,[8] it was reported that optic neuropathy was the most common manifestation of neurosarcoidosis, typically presenting with optic disc edema and severe visual loss.

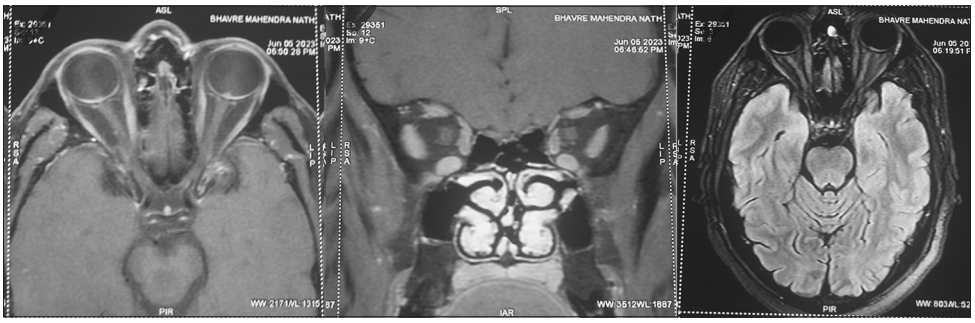

Figure 5: Magnetic resonance imaging brain with orbit with contrast was advised which showed no evidence of any abnormal lesion.

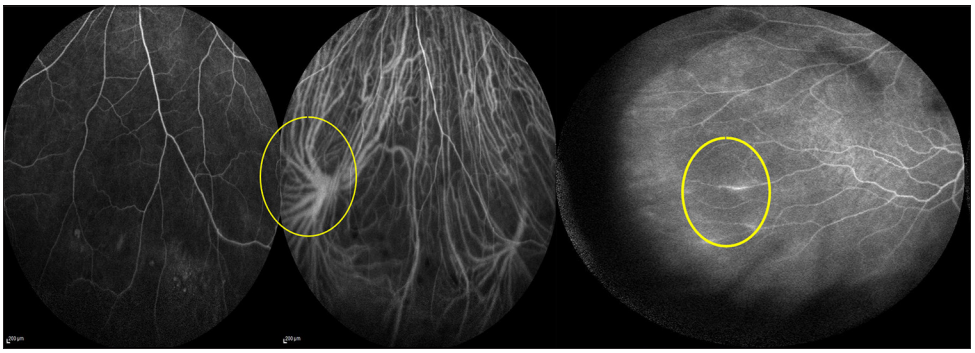

Figure 6: Fluorescein angiography showed multiple focal areas of pinpoint hyperfluorescence (yellow circles) as well as multiple small choroidal granulomas at posterior pole came into picture with indocyanine green angiography.

CONCLUSION

Our case of suspected sarcoidosis in a patient with a previous diagnosis of NAAION highlights the importance of detailed clinical examination with appropriate investigation in such cases. Early identification of patients is necessary because the delay in diagnosis hampers response to treatment. Identification of widespread disease warrants prolonged, often high-dose treatment with steroids, immunosuppressive drugs, and modern biological therapies.

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