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CASE REPORT

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Sarcoidosis presenting as optic neuritis with vision loss

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Abstract

Neurosarcoidosis is a rare complication of sarcoidosis and unusually presents as optic neuritis. We present the case of a 51-year-old man who complained of right vision loss. Brain magnetic resonance imaging showed asymmetrical enlargement of the right optic nerve. Chest computed tomography detected mediastinal and hilar lymphadenopathy. There were cutaneous nodules on the back. Biopsy of the mediastinal lymph node by endobronchial ultrasound-guided transbronchial needle aspiration and the skin showed noncaseating granulomas consistent with sarcoidosis. Serum angiotensin-converting enzyme level was elevated (34.2 IU/L) (normal: 8.3–21.4 IU/L). Based on these findings, he was diagnosed as neurosarcoidosis with optic neuritis. He was started on 1000-mg/day methylprednisolone intravenously for 3 days, followed by oral 50-mg/day prednisolone, which was gradually tapered for 8 weeks. Thereafter, the skin nodules and lymphadenopathy decreased and the right vision partially improved. Based on this rare case, sarcoidosis should be considered as a differential diagnosis in cases of optic neuritis.

KEYWORDS
endobronchial ultrasound-guided transbronchial needle aspiration, optic neuritis, sarcoidosis

INTRODUCTION

Sarcoidosis is a multisystemic granulomatous disease of unknown cause and is frequently localized in the lung, mediastinal/hilar lymph nodes, skin, liver, and eyes. Eye complications are common in sarcoidosis, with approximately 30%–50% of patients developing uveitis.[1] Uveitis is the most common ocular inflammatory response in patients with sarcoidosis; however, involvement of the optic nerve has been reported in only 1%–5% of patients with sarcoidosis.[2] Sarcoidosis that involves the optic nerve can cause progressive visual impairment and blindness. Here we present a case of sarcoidosis that presented with optic neuritis and with systemic disease in the mediastinal/hilar lymph nodes and skin.

CASE REPORT

A 51-year-old Japanese man visited the department of ophthalmology of our hospital with a two-month history of right vision loss without ophthalmalgia. He had a history of type 2 diabetes diagnosed 1 year ago, which had been controlled with oral medication without diabetic retinopathy. The patient was a former smoker (60 pack years) with no family history of sarcoidosis.

On physical examination, there were cutaneous nodules on the back. On neurologic examination, the patient was unremarkable, including the level of consciousness, mood, speech function, cranial-nerve function, muscle strength, sensory examination, reflexes, coordination, and gait. Visual field testing showed slight expansion of the right Marriotte blind spot. There was right optic nerve edema and haemorrhages on colour fundus photograph (Figure 1A) and optical coherence tomography (Figure 1B). The decimal best-corrected visual activity (BCVA) was 20/25 in the right eye. There was no uveitis to specifically suggest sarcoidosis.

Axial T1 weighted contrast magnetic resonance imaging (MRI) demonstrated the enhancement of the right optic nerve (Figure 1C). Further, in Fluid-attenuated inversion recovery image, there were no foci of demyelination in the brain that commonly appear in the corpus callosum and periventricular white matter, associated with multiple

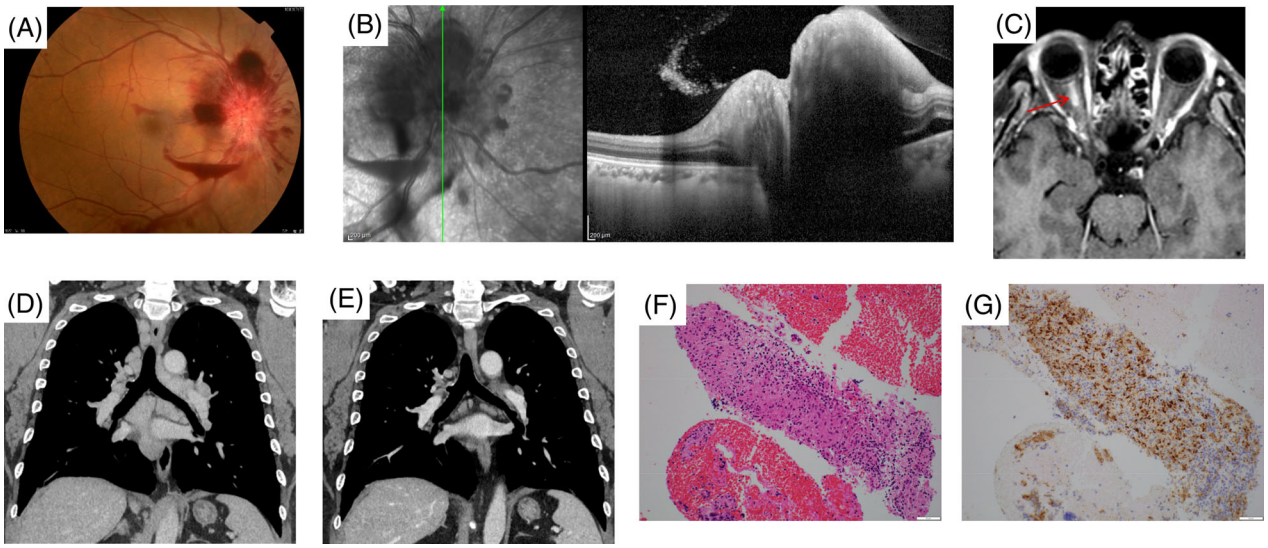

FIGURE 1 (A) Colour fundus photograph of the right eye shows edema of the optic disc and disc haemorrhage and partial vitreous haemorrhage. (B) Optical coherence tomography of the right eye demonstrates edema of the optic disc with haemorrhage. (C) Enhanced axial short inversion time inversion recovery image shows enhancement of the intraorbital right optic nerve (arrow). Coronal contrast-enhanced chest computed tomography shows (D) swollen hilar and mediastinal lymph nodes, which (E) improved after prednisolone treatment. (F) endobronchial ultrasound-guided transbronchial needle aspiration of the mediastinal lymph node shows nonnecrotizing granuloma. (Haematoxylin–Eosin staining, ×200); (G) immunohistochemistry shows infiltrating CD68-positive macrophages in the granuloma (×200)

sclerosis. Cerebrospinal fluid (CSF) study showed neither oligoclonal bands nor elevation of the IgG index. Slightly elevated levels of protein (74 mg/dL) (normal: 10–40 mg/dL), mononuclear cells (19 /3 μL) (normal: 1–5 /3 μL), and glucose (77 mg/dL) (normal: 40–70 mg/dL) were revealed by CSF studies. CSF bacterial, viral, and fungal cultures were negative. CSF cytology was negative. Laboratory examination showed increased levels of angiotensin-converting enzyme (ACE) at 34.2 IU/L (normal: 8.3–21.4 IU/L). Serum calcium level was normal (9.5 mg/dL) (normal: 8.5–10.2 mg/dL). Antinuclear antibody, serum interleukin-2 (IL-2) receptor, immunoglobulin G4, antineutrophil cytoplasmic antibody, T-spot, and aquaporin-4 antibody were negative. These findings suggested the diagnosis of sarcoidosis.

Contrast-enhanced chest computed tomography (CT) revealed mediastinal and hilar lymphadenopathy (Figure 1D). He underwent diagnostic bronchoscopy for bronchoalveolar lavage (BAL) and endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) of the mediastinal lymphadenopathies.[3] Forty-five mililiter of BAL fluid (left B4) was retrieved which contained 47% macrophages (normal: 75–95%), 16% neutrophils (normal: <3.0%) and 34% lymphocytes (normal: 4%–25%) and a slightly increased CD4/CD8 ratio of 1.96 (reference range: 1.2–1.8). EBUS-TBNA samples from the mediastinal lymph nodes showed noncaseating epithelioid cell granulomas with CD68-positive macrophage infiltration (Figure 1F, G).[4] Mycobacteria and fungal organisms were not detected. Biopsy from the cutaneous nodules also showed noncaseating epithelioid cell granulomas involving the cutaneous tissues.

Based on these findings, a diagnosis of sarcoidosis with optic neuritis was made. He was treated with intravenous methylprednisolone (1000 mg/day) for 3 days, followed by oral prednisolone at 50 mg/day. During tapering of the oral prednisolone to 20 mg at 8 weeks, the hilar and mediastinal lymph nodes decreased in size (Figure 1E), and the skin nodules resolved. Furthermore, there was resolution of the right optic haemorrhages, along with improvement of optic nerve edema and right vision loss. The right BCVA improved to 20/20.

DISCUSSION

We present a case of sarcoidosis complicated by optic neuritis. In sarcoidosis, the eye is the second most frequently affected organ after the lungs.[1] Uveitis is the most common ocular inflammatory response in sarcoidosis, and optic neuritis is extremely rare.[2] The different described mechanisms of vision loss secondary to optic nerve involvement have included intrinsic granulomatous infiltration, extrinsic compression, or intracranial hypertension involving the optic nerve, optic tracts, or the chiasm.[5] Optic nerve involvement in sarcoidosis is associated with poor visual outcome.[6] In this case, although the first clinical diagnosis for his vision loss was optic neuritis, its cause was unknown. The presence of mediastinal and hilar lymphadenopathy, subcutaneous nodules on the back, and elevated serum ACE raised the suspicion of sarcoidosis. The presence of noncaseating granulomas on pathologic evaluation of the mediastinal lymph nodes and skin nodules were supportive of a diagnosis of sarcoidosis.

Isolated involvement of the optic nerve without systemic manifestations is insufficient to establish a diagnosis of sarcoidosis.[5] Optic nerve sarcoidosis mimics the clinical and radiologic findings in optic nerve meningioma or glioma.[5] In this case, EBUS-TBNA was done to evaluate the mediastinal and hilar lymphadenopathy, because it is less invasive than mediastinoscopy and surgical biopsy. Treatment with prednisolone resulted in significant improvement of the lymphadenopathy and skin nodules on the back. Based on this case of sarcoidosis presenting as optic neuritis with vision loss, encounter of future cases with optic neuritis should prompt ophthalmologists to consider consultation with other specialists, such as neurologists and pulmonologists, for systemic investigation, including chest X-ray, chest CT scan, and serum tests for ACE. Sarcoidosis should be considered as a differential diagnosis in cases of optic neuritis with mediastinal and bilateral hilar lymphadenopathy.

AUTHOR CONTRIBUTIONS

Ryosuke Kataoka and Yoshiro Kai wrote the manuscript. All authors contributed to editing the manuscript and approved the final version of the manuscript.

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CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

All data generated or analysed during this study are included in this article.Further enquiries can be directed to the corresponding author.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained from the patient for publication of this case report and its accompanying images.

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