A rare manifestation of neuro-ophthalmic sarcoidosis: A case report

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Abstract

Purpose: Anterior uveitis is the most common ocular manifestation of sarcoidosis. Ocular involvement affects approximately 30–60% of patients with systemic sarcoidosis; however, optic disc edema is a rare event. We report a patient who presented with a rare case of sarcoidosis with neuro-ophthalmic manifestations.

Case report: A 22-year-old man was referred to our clinic with the primary complaint of a visual field defect over the temporal side of his right eye of 2 months duration. He did not have a history of systemic disease. At the first ophthalmic examination, the visual acuity, intraocular pressure, and slit lamp examination were normal. The fundus examination revealed bilateral optic disc edema. He was initially suspected of having a choroidal lesion between the disc and fovea of the right eye. To evaluate the possible lesion, the patient underwent brain magnetic resonance imaging (MRI), chest radiography, and chest computed tomography (CT). There were no abnormalities on the brain MRI, but the chest radiographs and CT images revealed bilateral mediastinal and hilar lymphadenomegaly. Histopathologic evaluation of an ultrasound-guided lymph node biopsy confirmed the diagnosis of sarcoidosis.

Conclusion: Neuro-ophthalmic manifestations of sarcoidosis are rare but may be the only presenting sign of an otherwise occult disease. A high clinical suspicion for sarcoidosis and its inclusion as a differential diagnosis are key to establishing the diagnosis and proper treatment.

1. Introduction

Sarcoidosis is a granulomatous disorder of unknown etiology with multisystemic and ocular manifestations. It occurs worldwide, but it is predominant in certain ethnic and racial groups; it is uncommon in people of Chinese descent. Major organs affected include the lungs, skin, eyes, liver, and lymph nodes. Ocular manifestations have been reported in 25–89.9% of sarcoidosis patients; however, posterior segment disease without anterior segment involvement is unusual. We report a rare case of sarcoidosis with posterior segment involvement as the only ocular manifestation in a Taiwanese man.

2. Case Report

In May 2012, a 22-year-old man presented to our clinic with a 2-month history of a visual field (VF) defect over the temporal side of his right eye. He did not have a history of systemic disease. At the first ophthalmic examination, the best corrected visual acuity (BCVA) of each eye was 20/20. Both pupils were 6 mm, round, and reactive to light without a relative afferent pupillary defect. The intraocular pressure was 11 mmHg in the right eye and 8 mm Hg in the left eye. There was no anterior uveitis or conjunctival granulomas in either eye. Funduscopic examination revealed bilateral swelling of the optic discs, a grayish-white mass in the superotemporal peripapillary region between the disc and fovea, and hard exudates around the fovea in the right eye (Fig. 1). Fluorescein angiography (FA) revealed optic disc staining in both eyes (Fig. 2). Automated static perimetry revealed a substantially enlarged blind spot of the right eye, which was compatible with the clinical symptom and fundus examination (Fig. 3).

The complete blood cell count and the serum biochemical profile were unremarkable, and a blood culture for bacterial and fungal pathogens was negative. Acid-fast stain and tuberculosis sputum culture were also negative. The serology test results were negative for human leukocyte antigen B27, antinuclear antibody, rheumatoid factor, reactive plasma reagin, human immunodeficiency virus antibodies, and toxoplasmosis antibodies. Immunoglobulin M antibody levels for cytomegalovirus herpes simplex virus, and varicella-zoster virus were normal. Magnetic resonance
Fig. 1. Optic disc edema in both eyes and a choroid lesion between the disc and fovea in the right eye.

Fig. 2. Fluorescein angiography shows optic disc staining in both eyes.
Fig. 3. Automated static perimetry reveals a substantially enlarged blind spot in the right eye.
Fig. 4. Chest computed tomography reveals multiple enlarged nodes in multiple compartments of the bilateral mediastinum and the bilateral hilar region (red arrows).

Fig. 5. (A) Granuloma tissue without caseous necrosis. (B) Cytokeratin staining is negative, which excludes a malignant tumor.

Fig. 6. After corticosteroid treatment, the volume of the peripapillary lesion decreased.
Fig. 7. After corticosteroid treatment, fluorescein angiography shows that bilateral optic disc staining resolved.
Fig. 8. After corticosteroid treatment, the visual field defect resolved.
imaging of the brain was performed with contrast. There was no space-occupying lesion in the brain or midline shift of the brain supratentorially or infratentorially. Radiographs and computed tomography (CT) images of the chest revealed multiple enlarged and confluent nodes (maximal dimension, 7 cm) at multiple compartments of the bilateral mediastinum, bilateral hilar region, and neck region. This finding was suggestive of sarcoidosis. The differential diagnosis included lymphoma (Fig. 4). Therefore, an ultrasound-guided biopsy of a cervical lymph node was performed. The histopathologic evaluation of the sample revealed granulomatous tissue without caseous necrosis, which stained negatively with cytokeratin (CK). Therefore, a diagnosis of lymphoma was excluded (Fig. 5). The patient was diagnosed as having neurosarcoidosis, based on the presentation, clinical testing, and biopsy results. Treatment was initiated with corticosteroids. After retrobulbar injection of triamcinolone 1.5 mL (10 mg/mL) into the right eye and systemic treatment with oral prednisolone (15 mg 3 times daily; total, 45 mg/d) for 3 months, the patient’s optic disc and macular lesions improved (Figs. 6 and 7). Approximately 1 month later, the VF defect resolved (Fig. 8).

3. Discussion

This case of sarcoidosis presented with bilateral optic disc edema, peripapillary choroidal granuloma, and a VF defect in a 22-year-old Taiwanese man. Sarcoidosis commonly affects young and middle-aged adults. It occurs worldwide and predominates in certain ethnic and racial groups such as African-Americans; it rarely occurs in Chinese patients.2 The most common presenting sign of ocular sarcoidosis is anterior uveitis,3 although choroidal granuloma and optic disc edema without anterior segment involvement has been rarely reported.4 The differential diagnosis for choroidal granuloma includes tuberculosis, sarcoidosis, metastatic tumor, and retinoblastoma. Histopathological proof with compatible clinical findings is the gold standard for a diagnosis. Conjunctival biopsy is a simple, safe, and specific diagnostic procedure for sarcoidosis,5 although there was no apparent conjunctival granuloma in our patient. An ultrasound-guided lymph node biopsy sample was collected instead. In a study by Sheu et al,6 biopsy samples were also primarily collected from nonocular tissues such as the lungs, skin, and peripheral lymph nodes. Corticosteroids are the mainstay of therapy in sarcoidosis patients.9 Treatment should be instituted when organ function is threatened.9 An international expert panel has suggested initiating treatment with oral prednisone at a daily dose of 20–40 mg.10 Our patient was followed up for 2 years after the initial institution. His VF improved after treatment, which included retrobulbar injection of triamcinolone 1.5 mL (10 mg/mL) in the right eye and a 3-month course of prednisolone (45 mg/d). There was no recurrence during the follow-up period. Neuro-ophthalmic manifestations of sarcoidosis are rare; however, they may be the presenting sign of the occult disease. Ophthalmologists should be aware of the ocular features of sarcoidosis to make an early diagnosis.4

Acknowledgments

The authors have no proprietary or commercial interests in any concept or product discussed in this article.

References

1. Herbort CP, Rao NA, Mochizuki M, members of Scientific Committee of First International Workshop on Ocular Sarcoidosis. International criteria for the diagnosis of ocular sarcoidosis: results of the first international Workshop on Ocular Sarcoidosis (IWOS). Ocul Immunol Inflamm. 2009;17:160–169.
2. Sheu SJ, Chang FP, Wu TT, Chuang CT. Ocular sarcoidosis in southern Taiwan. Ocul Immunol Inflamm. 2010;18:152–157.
3. Frohman L, Grigorian R, Slamovits T. Evolution of sarcoid granulomas of the retina. Am J Ophthalmol. 2001;131:661–662.
4. Augustin AJ, Boker T, Seewald S, Klassen PM. Solitary retinal granuloma as a presenting sign of sarcoidosis. German J Ophthalmol. 1994;3:71–72.
5. Chung YM, Lin YC, Huang DF, Hwang DK, Ho DM. Conjunctival biopsy in sarcoidosis. J Chin Med Assoc. 2006;10:472–477.
6. Iannuzzi MG, Rybicki BA, Teirstein AS. Sarcoidosis. N Engl J Med. 2007;357:2153–2165.