Abdominal sarcoidosis presenting as bilateral simultaneous optic disc granulomas

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ABSTRACT

A 24-year-old male presents with diminution of vision in both eyes of acute onset with floaters. He has a history of fever of unknown origin. Examination revealed bilateral optic disc granulomas with mild vitritis. Serum angiotensin converting enzyme was found to be elevated and tuberculin skin test was negative. Computed tomography scan of the chest showed clear lung fields with no hilar lymphadenopathy but mildly enlarged pretracheal lymph nodes. Computed tomography scan of the abdomen revealed multiple enlarged abdominal lymph nodes with hepatosplenomegaly, and ultrasound-guided biopsy of one of these lymph nodes showed chronic granulomatous inflammation consistent with sarcoidosis. Immunosuppressive therapy resulted in resolution of ocular inflammation with no recurrence.

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Introduction

Sarcoidosis is an idiopathic, multisystemic, granulomatous disorder characterized by formation of non-caseating granulomas in multiple organs most commonly the lungs. Ocular involvement is common, and an ocular complaint is often the first presenting symptom, occurring in about 20–30% of patients with sarcoidosis.¹

Neuro-ophthalmic manifestations of sarcoidosis are variable and less common than other ocular manifestations. They include optic neuropathy, cranial neuropathy, Horner's syndrome, optic chiasm, and optic tract involvement.^{1,2} Neuro-ophthalmic sarcoidosis has been reported to be associated with a lower incidence of pulmonary involvement as compared to other forms of sarcoidosis.²

Abdominal involvement by sarcoidosis is a relatively common finding on abdominal computed tomography of patients with biopsy-proven sarcoidosis. Manifestations include hepatomegaly, splenomegaly, hepatic and splenic nodules, and lymphadenopathy. These manifestations do not seem to correlate with the degree of pulmonary involvement.³

We report the findings of a patient who presented with bilateral simultaneous optic disc granulomas and was found to have abdominal sarcoidosis with no radiological evidence of pulmonary involvement. To the best of our knowledge, this is the first report of such a case.

Case report

A 24-year-old male presents with diminution of vision in both eyes of acute onset and progressive course for 1 week. He also complained of floaters. He has a history of fever of unknown origin for 1 month. Examination reveals a corrected-distance visual acuity of 20/80 in both eyes. Colour vision testing using the Ishihara cards was 12/12 in both eyes. Pupils were regular with a mildly sluggish response to light bilaterally. External ocular and anterior segment examination were normal. Posterior segment examination of both eyes revealed mild vitritis, dilated tortuous retinal veins, and bilateral hyperemic optic discs with nodular elevation suggestive of optic disc granulomas (Figure 1A and B). Fluorescein angiography was performed and revealed bilateral hyperfluorescent optic discs with leakage (Figure 1C and D). Laboratory investigations were done and showed elevated serum angiotensin

Figure 1. Fundus photography showing mild vitritis, dilated tortuous retinal veins, and bilateral swollen optic discs with nodular elevation suggestive of optic disc granulomas (A and B). Fluorescein angiography showing bilateral hyperfluorescent optic discs with leakage (C and D).

converting enzyme level at 84.1 U/L (8–52 U/L) and negative tuberculin skin test. Chest X-ray did not reveal any abnormality. Computed tomography (CT) scan of the chest showed clear lung fields with no hilar lymphadenopathy but mildly enlarged pre-tracheal and retrocaval lymph nodes. CT scan of the abdomen revealed hepatosplenomegaly with multiple enlarged abdominal lymph nodes including celiac, peri-pancreatic, porta hepatis, porto-caval, and para-aortic lymph nodes, the largest measuring about 4.2×2.7 cm. An ultrasound-guided core biopsy of an abdominal lymph node was done and showed chronic granulomatous inflammation with epithelioid and multinucleated giant cells with no evidence of caseation or malignancy. A diagnosis of sarcoidosis with bilateral optic disc granulomas was made and the patient was started on 40 mg/day prednisolone and 100 mg/day azathioprine. Two months following treatment, corrected-distance visual acuity improved to 20/40 in both eyes, posterior segment examination revealed resolution of the disc swelling and granulomas bilaterally (Figure 2A and B), and fluorescein angiography showed no disc hyperfluorescence or leakage (Figure 2C and D). Gradual tapering of prednisolone dose was done, and the patient was maintained on 10 mg/day of

prednisolone and 50 mg/day of azathioprine for 1 year with no recurrence.

Discussion

The anterior visual pathway is estimated to be affected in about 1–5% of patients with sarcoidosis, with chiasmal involvement being more common than intraorbital involvement.⁴ Optic nerve involvement occurs in several forms and includes optic neuritis, perineuritis, papilledema, and optic nerve granulomas.^{5–7} Infiltration of the optic nerve head by sarcoid granulomas may resemble papillitis but appears clinically as optic disc nodules, which requires histopathology for confirmation.^{5,7} This was suspected in our case due to the nodular elevation of the optic nerve heads. Few cases of sarcoidosis that presented as simultaneous bilateral optic nerve involvement have been reported in the literature and the diagnosis is usually challenging in these cases.^{6–8} The presence of posterior segment manifestations, such as vitritis in the presented case, may assist in the diagnosis.⁷

Abdominal involvement by sarcoidosis is not uncommon. In a study of abdominal CT findings in

Figure 2. Fundus photography showing resolution of optic disc swelling and granulomas bilaterally with no residual vitritis (A and B). Fluorescein angiography showing no evidence of disc hyperfluorescence or leakage (C and D).

59 patients with biopsy-proven sarcoidosis, 31% had abdominal lymphadenopathy with porta hepatis and para-aortic lymph nodes being most commonly affected. The liver and spleen were enlarged in 29% and 33%, respectively. There was no relation detected between chest X-ray appearance and abdominal findings as in our patient. No data, however, regarding ocular involvement was reported in this study.³ This data, together with the presented case, suggest that abdominal manifestations of sarcoidosis may be helpful, and in fact sometimes the major clue, in the diagnosis of systemic sarcoidosis. This is particularly true in case of absence of chest manifestations.

Diagnosis of sarcoidosis can be suspected by clinical, laboratory, and radiological findings typical of sarcoidosis and by excluding other possible conditions. Serum angiotensin converting enzyme for example is elevated in 75% of patients with sarcoidosis and may be helpful in the diagnosis as in our case. Confirming the diagnosis of sarcoidosis, however, depends on histopathological demonstration of non-caseating granulomas and should be done in all cases.⁹

Corticosteroids are the mainstay of treatment for sarcoidosis. Because long-term therapy with corticosteroids is usually needed to maintain remission and prevent recurrent inflammation, an immunomodulatory agent such as azathioprine can be used to decrease corticosteroid dependence and improve treatment tolerance.¹

In summary, we report a patient who presented with bilateral optic disc granulomas and was found to have biopsy-proven abdominal sarcoidosis with no radiological evidence of pulmonary involvement who responded well to immunosuppressive treatment.

Ethical approval

This report was approved by Cairo University Research Ethics Committee and followed the tenets of the Declaration of Helsinki.

Declaration of interest

The authors declare that there are no conflicts of interest. The authors alone are responsible for the writing and content of the article.

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References

1. Pasadhika S, Rosenbaum JT. Ocular sarcoidosis. *Clin Chest Med.* 2015;36:669–683. doi:10.1016/j.ccm.2015.08.009.
2. Koczman JJ, Rouleau J, Gaunt M, Kardon RH, Wall M, Lee AG. Neuro-ophthalmic sarcoidosis: the University of Iowa experience. *Semin Ophthalmol.* 2008;23(3):157–168. doi:10.1080/08820530802007382.
3. Warshauer DM, Dumbleton SA, Molina PL, Yankaskas BC, Parker LA, Woosley JT. Abdominal CT findings in sarcoidosis: radiologic and clinical correlation. *Radiology.* 1994;192:93–98. doi:10.1148/radiology.192.1.8208972.
4. Engelken JD, Yuh WT, Carter KD, Nerad JA. Optic nerve sarcoidosis: MR findings. *AJNR Am J Neuroradiol.* 1992;13:228–230.
5. Jampol LM, Woofin W, McLean EB. Optic nerve sarcoidosis: report of a case. *Arch Ophthalmol.* 1972;87:355–360. doi:10.1001/archophth.1972.01000020357023.
6. Kidd DP, Burton BJ, Graham EM, Plant GT. Optic neuropathy associated with systemic sarcoidosis. *Neurol Neuroimmunol Neuroinflamm.* 2016;3:e270. doi:10.1212/NXI.0000000000000270.
7. Beck AD, Newman NJ, Grossniklaus HE, Galetta SL, Kramer TR. Optic nerve enlargement and chronic visual loss. *Surv Ophthalmol* 1994;38(6):555–566. doi:10.1016/0039-6257(94)90148-1.
8. Cem Y, Basar A, Ozlem O, Nilgul Y, Selcuk P. Primary bilateral optic nerve sarcoidosis. *Neuroophthalmol.* 2005;29(4):165–172. doi:10.1080/01658100500218038.
9. Heinle R, Chang C. Diagnostic criteria for sarcoidosis. *Autoimmun Rev.* 2014;13(4–5):383–387. doi:10.1016/j.autrev.2014.01.035.