Case report

Sarcoidosis presenting as Wallenberg syndrome and panuveitis

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A B S T R A C T

Sarcoidosis is a multi-system disease with neurological involvement being one of the more rare manifestations. We report a case of a patient who presented with the lateral medullary syndrome and panuveitis as her initial manifestation of sarcoidosis. The patient's course was further complicated by renal involvement. Lacrimal gland and renal biopsies showed non-caseating granulomas without evidence of infection, establishing the diagnosis. Intracranial vertebral artery involvement was confirmed by brain imaging. Bilateral hilar lymphadenopathy with upper lobe predominant nodules on chest imaging was consistent with asymptomatic pulmonary involvement. Systemic steroid therapy is indicated for treatment of ocular sarcoidosis, with standard stroke management indicated for the treatment of lateral medullary syndrome.

1. Introduction

Sarcoidosis is a multi-system disease of unclear etiology, but is most likely related to an antigen driven process that is yet to be identified. The initial presentation of sarcoidosis varies greatly, but neurosarcoidosis is one of the more rarely seen initial manifestations. While various different neurological patterns have been described in association with sarcoidosis, lateral medullary syndrome (Wallenberg syndrome) in concurrence with panuveitis is a new entity we describe in our case report.

2. Case report

A 58-year-old African American woman presented with a one-week history of blurry vision and right eye pain. She reported bilateral ocular puffiness, insomnia, dry skin, and weight loss over one month. She denied dyspnea, fever, chin erythema or joint pain.

On initial physical exam the patient was noted to be hypertensive with a regular heart rate without any murmurs, and clear lungs. She had reduced visual acuity bilaterally with reactive pupils, and small amplitude bilateral rotary nystagmus. Fundal examination showed bilateral vitritis, and multiple intra-retinal hemorrhages with vascular sheathing and leakage on fluorescein angiography (Fig. 1), consistent with retinal vasculitis.

Within a few hours of admission, the patient endorsed a severe headache, left facial numbness, vomiting, and dizziness. On repeat physical exam she had left tongue deviation, left upper extremity drift, ataxia, and dysmetria with finger-to-nose test. Sensory exam showed loss of pain and temperature sensation to the left side of her face, and in the right upper and lower extremities; deep tendon reflexes were intact. The patient also developed dysphagia, which was confirmed by a speech pathologist. The patient underwent emergent Computed Tomography (CT) brain imaging, which did not show any abnormalities. Magnetic Resonance Imaging (MRI) of the brain showed an area of restricted signal intensity in the left lateral medulla (Fig. 2A) with ischemic changes also noted along the left and right sides of the mid pons (Fig. 2B). Trans-thoracic echocardiography did not show signs of a cardio-embolic source for the patient's ischemic stroke.

Given the acute ocular findings and the confirmed ischemic stroke the patient was initiated on systemic prednisone, topical ocular prednisolone acetate, and cycloplegic drops. CT of the chest showed intrathoracic lymphadenopathy with peri-bronchovascular micronodules in the upper lobes, consistent with sarcoidosis (representative slice, Fig. 3). The patient had elevated levels of homocysteine, angiotensin converting enzyme (ACE), lysozyme, and lactate dehydrogenase. A lacrimal gland biopsy showed non-caseating granulomas (Fig. 4A) with

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negative microbiology stains.

The patient improved significantly on systemic steroids with resolution of ocular symptoms and was successfully discharged to a subacute rehabilitation facility for further management of ataxia and dysphagia related to her ischemic stroke. Her post hospital course has been complicated by avascular necrosis with subsequent bilateral hip decompression and bone grafts, frequent uveitis recurrences requiring further steroids and steroid sparing immunologic agents, as well as hematochezia requiring endoscopy and colonoscopy. On recent follow up the patient was noted to have deteriorating kidney function, which led to a renal biopsy that showed non-caseating granulomatous infiltration with negative microbiology stains (Fig. 4B).

3. Discussion

Our patient’s initial presentation was consistent with neurosarcoidosis, more specifically lateral medullary syndrome (Wallenberg syndrome) and panuveitis. Five to ten percent of individuals with known sarcoidosis have neurologic complications, and about 50% of patients initially present with neurologic symptoms at the same time as the initial diagnosis of sarcoidosis [1]. About 33% of patients with neurosarcoidosis have more than one simultaneous neurologic manifestation [2]. To our knowledge our patient’s presentation with panuveitis and the Wallenberg syndrome is the first such described case.

Our patient presented with an ischemic stroke in the posterior circulation. Lateral medullary infarction is the most common presentation of intracranial vertebral artery occlusion. The syndrome involves the vestibulo-cerebellar apparatus, the sensory and bulbar tracts, and respiratory dysfunction [3]. Collectively all of these affected brain territories lead to dizziness, nystagmus, emesis, ataxia, ipsilateral face and
contralateral trunk pain and temperature sensory loss, dysphagia, and diaphragmatic dysfunction. All of these signs, excluding respiratory dysfunction, were present in our patient. In a case series of 54 patients with at least a possible diagnosis of neurosarcoidosis, 24% presented with optic neuropathy, 19% presented with myelopathy, and 17% presented with seizures or headaches [4]. There have been case reports of neurosarcoidosis presenting as ischemic stroke, but none with the lateral medullary syndrome [5–11].

One fifth of patients develop uveitis as an initial presentation of sarcoidosis, and it is frequently associated with retinal vasculitis. Panuveitis is the simultaneous inflammation of the anterior chamber, vitreous humor, and the retina. Panuveitis can be associated with systemic disorders such as inflammatory diseases and infections, or be an isolated entity [12]. A study of 83 patients with biopsy proven sarcoidosis-associated uveitis showed that any elevated enzyme test, such as ACE and lysozyme, and suggestive chest imaging identified 100% of sarcoidosis related uveitis cases [13].

The determination of whether to treat sarcoidosis depends on the organs involved, with cardiac, ocular, and neurological involvement almost always requiring treatment. The treatment of Wallenberg syndrome follows the standard of care for all ischemic strokes, including aggressive physical rehabilitation and secondary prophylaxis with aspirin and statin. Panuveitis is a steroid responsive disease, but is recurrent and is associated with multiple steroid related complications.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.rmcr.2018.03.002.

References


Fig. 4. (A) Right lacrimal gland biopsy, Hematoxylin and Eosin (H&E) Stain, 100x; (B) Renal Biopsy, Hematoxylin and Eosin (H&E) Stain, 100x. Both show granulomas (black arrows) composed of tightly-packed histiocytes with multinucleate forms, and a surrounding chronic inflammatory infiltrate.