

# Retrobulbar Optic Neuropathy As Initial Presentation Of Sarcoidosis

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## Abstract

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. Even though Sarcoidosis manifests primarily as lung disease, it can involve any organ system to varying extent. Involvement of the nervous system is relatively rare and occurs in less than 5% of patients. Sarcoidosis can affect different parts of the nervous system, with cranial nerve involvement being more frequent. Neurosarcoidosis is a diagnostic challenge, especially when systemic manifestations of sarcoidosis are absent. We present an isolated case of retrobulbar optic nerve sarcoidosis presenting as initial manifestation of the disease in a 45 year old otherwise healthy female.

## CASE REPORT

We present a case of 45 years old African American female referred by her ophthalmologist to neurology clinic. The patient presented first for difficulty of vision involving the left eye. She described it as constant blurring and decreased visual acuity associated with moderate to severe pain. Onset was gradual in few weeks. The symptoms were not alleviated or worsened by any known factor. She was apparently in a good health without any significant medical problems prior to presentation. Rest of the review of systems was negative. This was treated by her ophthalmologist with prednisone with assumption of optic neuritis. Moderate improvement of symptoms was noted. Attempts to taper steroid, however, were associated with relapse of symptoms. Ophthalmic exam showed sharp disc bilaterally with normal cup and disc size. Retinal exam was normal. Pupils were equal and reactive with normal size. No Marcus Gun pupil. Visual acuity was 20/30 on right eye and 20/70 on left eye. Extra ocular movement was normal. Cranial and neurological exam was within normal limit without any localizing findings. Other physical findings were benign. Subsequent studies including Complete Blood Count, Chemistry profile, Erythrocyte Sedimentation Rate, Venereal Disease Research Laboratory, Antineutrophilic Antibody profile, Vit B12, Human Immunodeficiency test, Cerebrospinal Fluid (CSF) analysis, Angiotensin Converting Enzyme level, Chest X ray, PPD skin test, were all normal. Visual Evoked Potential however showed abnormality with delayed latency on left eye. MRI of brain and orbits with and

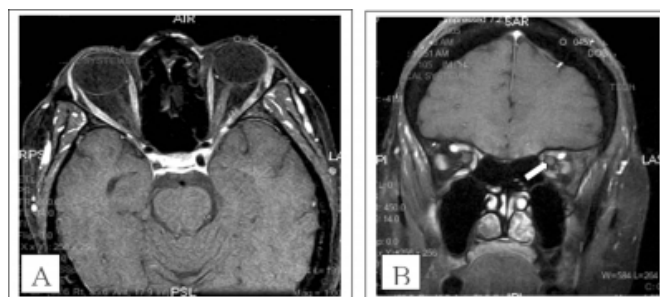
without contrast showed enhancement of left optic nerve (Figure 1). Gallium scan of the lung showed strong radiotracer uptake.

Considering patient's age, gender, race, response of symptoms to steroids, MRI finding, and positive Gallium radiotracer lung uptake; a diagnosis of optic nerve sarcoidosis involving retrobulbar part was made.

She was subsequently referred to pulmonary clinic for bronchoscope and tissue diagnosis. Pathology result was inconclusive from bronchial tissues and mildly enlarged hilar lymph nodes despite positive Gallium tracer uptake. Tissue culture for tuberculosis and fungal analysis were negative. She is on daily 10mg dose of prednisone at this time. She is relatively pain free and her visual acuity on left eye has increased from 20/70 to 20/40.

## Figure 1

Figure 1: MRI Showing Contrast Enhancement of Left Optic Nerve in Transverse (A) and Coronal (B) Views (arrows).



## **DISCUSSION**

Sarcoidosis is a multisystem granulomatous disease of unknown etiology<sup>1, 2</sup>. Neurosarcoidosis is atypical manifestation of sarcoidosis occurring in about 5% of cases of systemic sarcoidosis<sup>3</sup>. Postmortem studies however suggested involvement may be even higher<sup>5</sup>.

Diagnosis is easier to make in the patients with already existing multisystem disease. However, if systemic manifestations are absent and neurologic involvement is the initial presentation the diagnostic process is arduous<sup>4</sup>.

Commonly seen syndromes in neurosarcoidosis include: granulomatous leptomeningitis with multiple cranial nerve involvement; local granulomatous mass lesion involving pituitary–hypothalamic area; sarcoid granuloma in the parenchyma of the brain; and optic nerve involvement. Involvement of optic nerve by sarcoid lesion was first reported in 1964<sup>3, 6</sup>. It is the most frequently affected cranial nerve next to the facial nerve<sup>4, 11</sup>. Sarcoid granuloma may involve optic nerve anywhere along its path from the nerve head to the chiasm. Granulomatous infiltration as well as compression was demonstrated by biopsy material<sup>9</sup>.

Clinically it can present as acute or chronic, painful or painless visual loss<sup>8</sup>. Asymptomatic involvement was also demonstrated by Visual Evoked Potential<sup>12</sup>.

Even though papillitis, optic disc atrophy, anterior segment involvement was mentioned in the literature since 1964, isolated retrobulbar involvement of the optic nerve was first reported by Rush in 1980<sup>7</sup>. He reported 27 years old black woman with visual symptoms and clear discs who was later diagnosed with retrobulbar sarcoidosis without systemic involvement.

In most of the reported cases the diagnosis was challenging as there were alternative differentials for the explanation, including multiple sclerosis. Our patient had normal CSF analysis with no oligoclonal bands or basic myelin protein. MRI didn't show additional lesions. Inflammatory markers were all normal. Positive Gallium scan of lungs speaks against other differentials mentioned, particularly if her age and ethnicity are also considered. Her symptoms were exquisitely responsive to steroid which is also characteristic

of the lesion as mentioned in previous case reports.

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