Radiological evolution and delayed resolution of an optic nerve tuberculoma: Challenges in diagnosis and treatment

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Abstract

Optic nerve tuberculomas are rarely reported and their natural history, prognosis, and duration of required treatment remain unclear. A 40-year-old immunocompetent male presented with complete loss of vision in his right eye, which had evolved over 6 weeks. He had optic atrophy on examination. Initial imaging showed right optic nerve swelling and thickening suggesting an infiltrative inflammatory optic neuropathy (infectious or noninfectious). Serial imaging revealed appearance of ring enhancement with a necrotic centre. Biopsy and culture of the coexistent parietal lobe lesion revealed Mycobacterium tuberculosis. Persistent optic nerve granuloma with evidence of radiological improvement was noted at 18 months follow-up with antituberculous therapy (ATT). Visual recovery could not be achieved. The salient features in this case include the clinical presentation initially mimicking an infiltrative or compressive optic neuropathy, rapid radiological evolution into a tuberculoma, subtle paradoxical radiological worsening after initiation of ATT and persistence of granuloma on follow up scan. The challenges involved in early diagnosis and during the treatment course will be discussed.

Key Words

Antituberculous therapy, granuloma, magnetic resonance imaging, optic nerve, tuberculoma

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Ann Indian Acad Neurol 2013;16:114-7

Case Report

A 40-year-old male presented with complaints of decreased vision in the right eye of 4 months duration. There was pain in the right eye at onset and the visual loss progressed to complete loss of vision within 6 weeks of onset of symptoms. There was no history of seizures or focal limb weakness. There were no systemic features like fever, loss of appetite, or weight. Previous treatment history for the visual loss included a short course of oral steroids; no improvement was noted with the same.

He presented to us nearly 4 months after onset of visual symptoms and nearly 10 weeks after manifesting complete visual loss. On examination, there was no light perception in the right eye. Relative afferent pupillary defect and optic disc pallor were noted. Magnetic resonance imaging (MRI) brain and orbit at this time (16 weeks after onset of symptoms) revealed a homogenously enhancing, diffusely thickened optic nerve [Figure 1a] with nodular enhancing lesions in the right parietal and frontal lobes. [Figures 2 a1 and a2]. Human immunodeficiency virus enzyme-linked immunosorbent assay (HIV ELISA) was negative. Cerebrospinal fluid (CSF) analysis including cultures was noncontributory. There was no significant lymphadenopathy. Chest X-ray and abdomen ultrasonography were noncontributary. As there was still significant enhancement of the optic nerve, possibility of an infiltrative optic neuropathy due to an infectious (tuberculosis, cysticercosis) or inflammatory condition (sarcoidosis) was considered most likely. Infiltrations from lymphomatosis, gliomatosis, and other compressive optic neuropathies were considered less likely especially considering the brain lesions. At this stage, he was asymptomatic for the brain lesions and had already completely lost his vision in the right eye. Initiation of empiric antituberculous therapy was strongly contemplated. However, considering the risk of missing other diagnosis in which different treatment will be required, it was decided to go ahead with a biopsy for definitive diagnosis. The options included an optic nerve biopsy versus brain biopsy.

While awaiting the biopsy in the ward, mild impairment of dexterity of the left hand was noted. Antituberculous therapy

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	DOI: 10.4103/0972-2327.107722

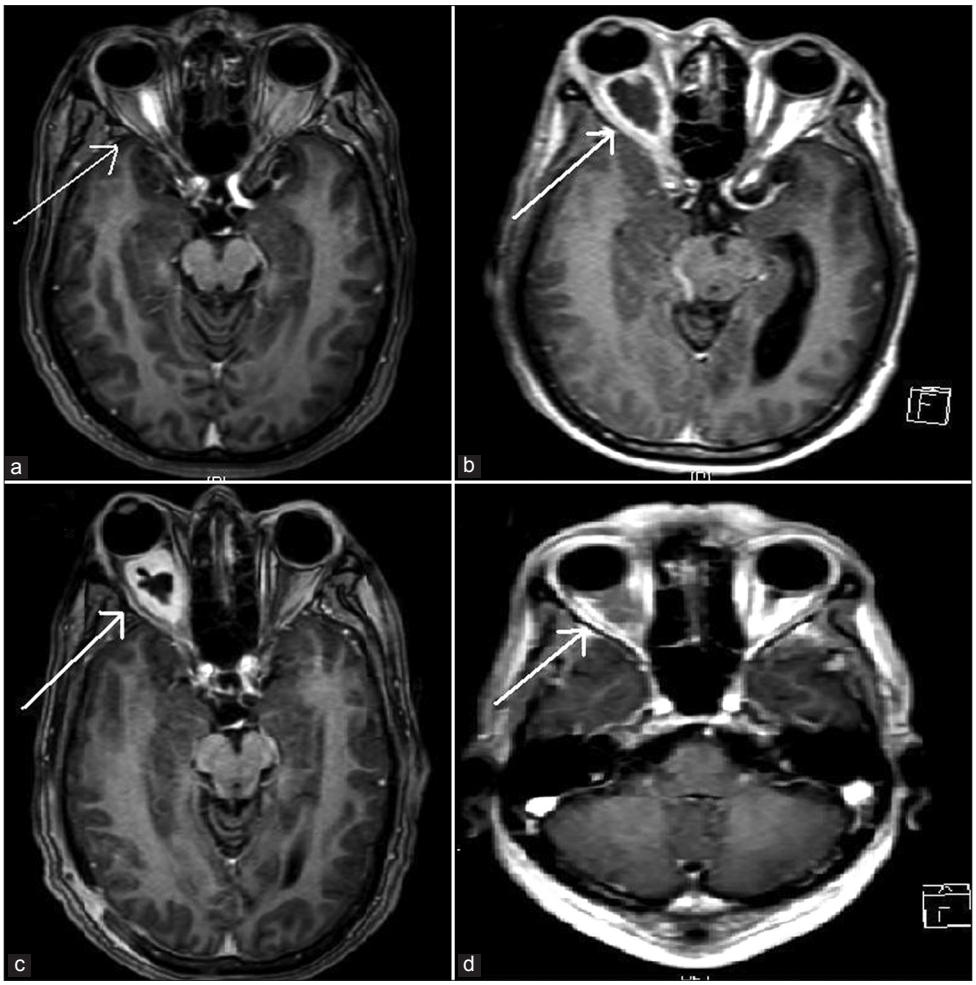

Figure 1: MRI (T1 W axial, postcontrast) showing sequential evolution of right optic nerve tuberculoma (a) diffuse thickening with homogenous enhancement (b) ring enhancement with proptosis (c) necrotic lesion with thicker enhancing rim (after 8 weeks of treatment) (d) reduction in size and degree of enhancement (18 months)

(ATT) (isoniazid, rifampicin, pyrazinamide, and ethambutol) with dexamethasone was started after a repeat imaging. A repeat MRI brain revealed conglomerate ring enhancing lesions in right frontal and parietal lobes with perilesional edema, mass effect, and midline shift. [Figure 2b] There was also a 30 × 21 mm ring enhancing lesion in the right optic nerve [Figure 1b] with associated thickening. Biopsy from the right parietal lesion revealed necrotizing granulomatous inflammation with numerous acid fast bacilli. Pansensitive mycobacterium tuberculosis was grown on culture. ATT with steroids was continued. Decompression of optic nerve lesion was not done as he already had complete loss of vision with optic atrophy. The mild weakness of the left hand improved completely. Repeat imaging at 2 months follow up revealed subtle increase in the size of the right optic nerve necrotic rim enhancing lesion [Figure 1c]. There was remarkable increase in the size, number, and perilesional edema of the brain lesions as well consistent with paradoxical worsening. [Figure 2c] The dosage of steroids was hiked up and ATT was continued. Repeat MRI at 18 months showed significant resolution of the intracranial lesions. [Figure 2d] The optic nerve lesion persisted, though there was a decrease in the size and degree of enhancement [Figure 1d]. Hence, ATT was continued.

Discussion

Tuberculosis can have a variety of ocular manifestations, which include anterior uveitis, posterior uveitis, panuveitis, retinitis, or endophthalmitis.[1] Visual compromise can also occur in tuberculosis due to hydrocephalus, optochiasmatic arachnoiditis, tuberculomas, optic neuropathy, and ethambutol toxicity. Tuberculomas of the optic nerve *per se* are rare.[2-4] Keshwachar reported a case of optic nerve tuberculoma proven histopathologically after enucleation.[2] Lana-Peixoto *et al*. reported an intrinsic tuberculoma of the left intracranial optic nerve on autopsy of a 1 1/2-year-old child with tuberculous meningitis and disseminated military tuberculosis.[3]

Optic neuropathy can occur from infectious infiltration or inflammation anywhere along the length of the optic nerve, and can present as disc edema or retrobulbar optic neuritis. This optic nerve involvement can occur from hematogenous spread or extension from the choroid. Differentiation from other inflammatory optic neuropathies may be initially difficult especially in the absence of systemic involvement. Contrary to intracranial tuberculomas, the natural history of optic nerve tuberculoma is unclear based on currently available literature.

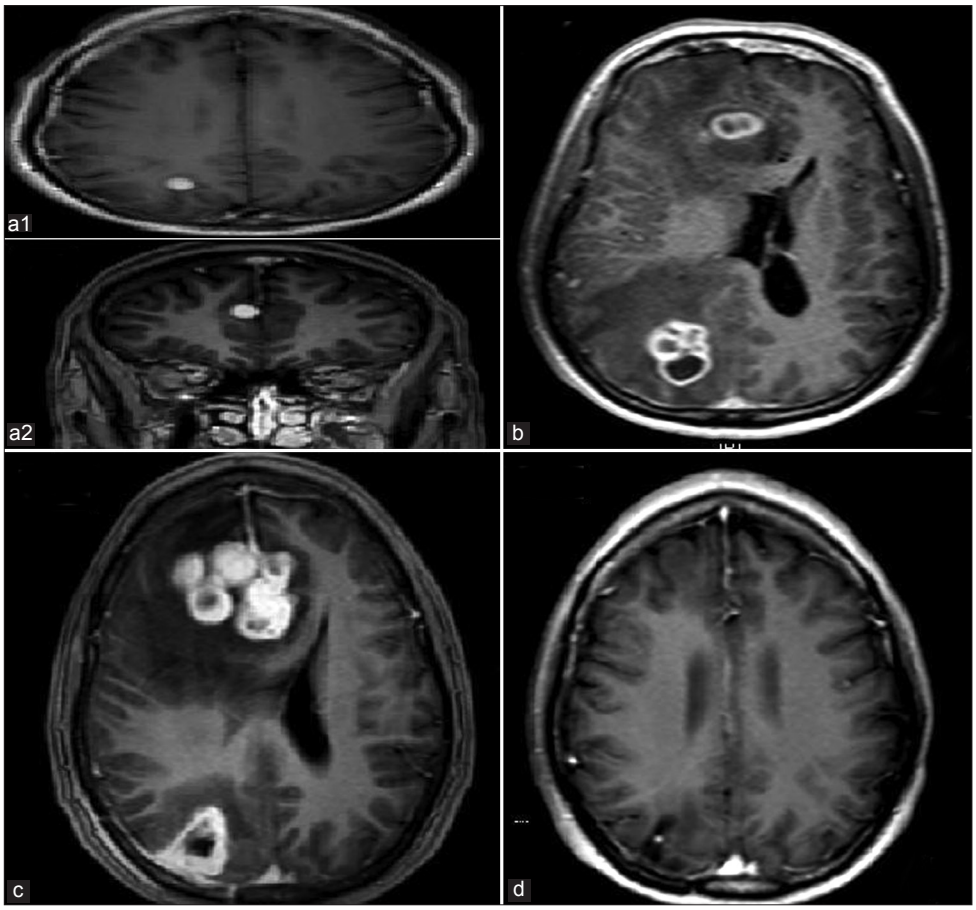

Figure 2: MRI brain (T1W axial, postcontrast) showing corresponding images from intracranial parenchymal lesions (a) nodular enhancing parietal lesion (A1) and frontal lesion (A2, coronal) (b) conglomerate ring enhancing lesions in right parietal and frontal lobes with mass effect (c) paradoxical worsening of these lesions (after 8 weeks of treatment) (d) significant resolution in brain lesions (18 months)

A high index of suspicion is warranted for this disease which can be difficult to diagnose and treat.

To the best of our knowledge, the spectrum and sequential evolution of radiological findings in an optic nerve tuberculoma has not yet been described in literature. Our patient had progressive visual deterioration, radiological worsening of optic nerve lesions evolving from nerve thickening to enlarging tuberculomas, subtle paradoxical radiological worsening of the optic nerve lesion, isolation of pansensitive mycobacteria on culture and radiologically resolving but persistent optic nerve granuloma at 18 months follow up.

The patient had only visual symptoms at onset in the form of progressive visual loss. The initial imaging in this case simulated an infiltrative optic neuropathy. Differentiation from other infectious or noninfectious inflammatory (sarcoidosis) optic neuropathies can be difficult at initial presentation especially if there are no parenchymal brain lesions. Extensive tuberculous optic nerve involvement with radiological appearance simulating an optic nerve glioma has also been reported.[5] Differential diagnosis for an optic nerve granuloma in immunocompromised hosts should also include toxoplasmosis, cryptococcosis, lymphoma, and less likely cytomegalovirus.[6]

Early diagnosis of tuberculous optic neuropathy can at times be challenging especially if the symptoms are restricted to vision involvement. Potential morbidities of an optic nerve exploration for biopsy include exacerbation of visual loss, CSF leaks and seeding of infection into the central nervous system (CNS). This case has been highlighted to describe the spectrum of radiological findings that can occur with optic nerve tuberculosis. Prompt recognition of the radiological patterns can potentially obviate the need for an invasive procedure like an optic nerve biopsy especially in patients who have some residual vision.

Presence of optochiasmatic tuberculomas and arachnoiditis has both therapeutic and prognostic implications in treatment. Paradoxical worsening due to immune reconstitution, release of new antigen targets with mycobacterial killing and hypersensitivity to these antigens can occur while on treatment and this needs to be carefully differentiated from treatment failure due to resistant mycobacteriae.[7] The risk factors for paradoxical worsening include disseminated tuberculosis, associated HIV infection, and infliximab treatment. There is a high load of bacilli in granulomatous lesions in these situations.[8] Isolation of drug sensitive mycobacteriae, favorable clinical response, increase in total lymphocyte count, and drug susceptibility assays (to identify ATT drug resistance) can be valuable in differentiating paradoxical worsening from treatment failure.[7] High dose

and prolonged corticosteroid treatment may be effective in preserving vision in cases of paradoxical worsening by reducing the host–organism reaction. Timely surgical decompression of the anterior visual pathways may also help in preserving vision when medical treatment fails.[9] Surgical decompression was not considered in our case as the patient already had complete visual loss prior to presentation with optic atrophy.

Similar to intracranial tuberculomas,[10] rate of resolution can be slow as seen in this case. Persistent optic nerve granulomas can be seen even after 18 months of ATT possibly implicating need for prolonged duration of treatment. Further case reports and series can give valuable information needed for appropriate management (duration of treatment) and prognostication of these patients. The need for early diagnosis with high index of suspicion, prompt initiation of therapy and anticipation and appropriate management of complications like paradoxical reactions has to be emphasized. Unfortunately, our patient as mentioned before had manifested complete loss of vision with optic atrophy prior to presentation and functional visual recovery could not be achieved even after initiation of ATT.

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How to cite this article: Sivadasan A, Alexander M, Mathew V, Mani S, Patil AB. Radiological evolution and delayed resolution of an optic nerve tuberculoma: Challenges in diagnosis and treatment. Ann Indian Acad Neurol 2013;16:114-7.
Received: 11-02-12, **Revised:** 08-03-12, **Accepted:** 10-06-12

Source of Support: Nil, **Conflict of Interest:** Nil