Unique case of midbrain tuberculoma presenting as isolated inferior rectus palsy with nystagmus

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Isolated brain stem tuberculoma constitutes about 5% of all intracranial tuberculomas. A case of isolated inferior rectus palsy with downbeat nystagmus due to presumed midbrain tuberculoma in an immunocompetent patient is described here. This report documents a rare entity of a combination of partial third nerve palsy with pupil involvement along with downbeat nystagmus.

Key words: Downbeat nystagmus, midbrain, nerve palsy, tuberculoma

Tuberculosis remains a significant health hazard in developing countries. Intracranial tuberculoma accounts for 5%–8% of space-occupying lesions in the brain.[1] They are usually located in the cerebral or cerebellar hemisphere due to high blood supply to these areas; brain stem is an uncommon location though. Isolated brain stem tuberculomas constitute only 5% of all intracranial tuberculomas. The usual presentations are isolated/multiple cranial nerve palsies, unilateral saccadic paralysis or one-and-a-half syndrome.[2]

We describe a case of isolated Inferior rectus palsy with downbeat nystagmus due to presumed midbrain tuberculoma in an immunocompetent patient. This report documents a rare entity – a combination of partial third nerve palsy with pupil involvement along with downbeat nystagmus.

Case Report

A 21-year-old girl presented with binocular diplopia and upward deviation of right eye for 2 days. She complained of vertical separation of images more so in downgaze and also had a history of 5 kg weight loss in the last 2 months. On examination, best-corrected visual acuity in both eyes was 6/6. She had right head tilt. On orthoptic evaluation, 10-prism diopter (PD) right hypertropia was noted for near and distance with the right head tilt. On prism cover test, right hypertropia measured 25 PD on the right gaze and 16 PD on the left head tilt confirming the Parks–Bielschowsky three-step test for right inferior rectus palsy. Right eye showed well dilated (6 mm) and fixed pupil, with absent response for both direct and consensual light reflex. Left eye anterior segment was within normal limits. Fundus was normal in both eyes. Examination of extraocular movements revealed bilateral, symmetrical, low amplitude and frequency, vertical nystagmus on downgaze with underaction of right inferior rectus muscle (dextrodepression) [Fig. 1]. Hess charting confirmed underaction of right inferior rectus [Fig. 2]. A definitive diagnosis of right pupil involving partial third nerve palsy with downbeat nystagmus was made. To rule out the cause for downbeat nystagmus and surgical reasons as a cause for pupil involvement in a case of third nerve palsy, magnetic resonance imaging (MRI) brain plain and contrast with MR angiography was ordered. It showed an

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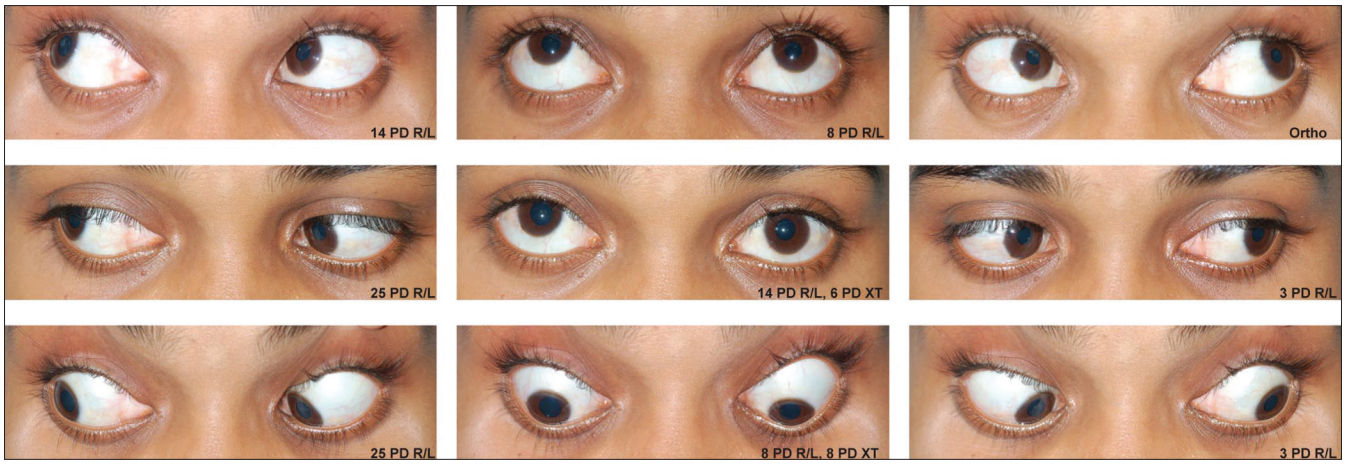

Figure 1: Extraocular movements showing restriction of dextrodepression

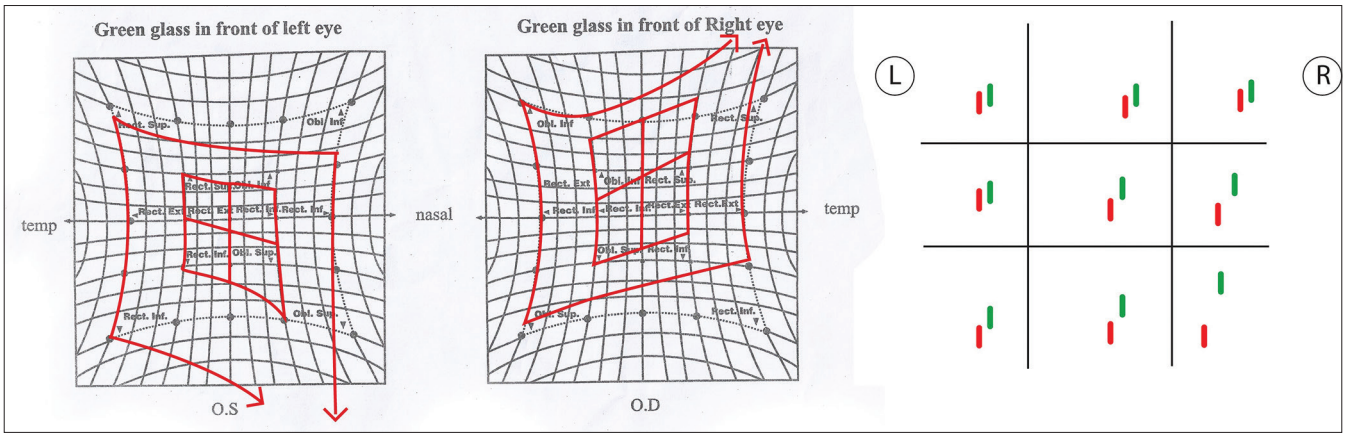

Figure 2: Hess and diplopia chart – right inferior rectus restriction

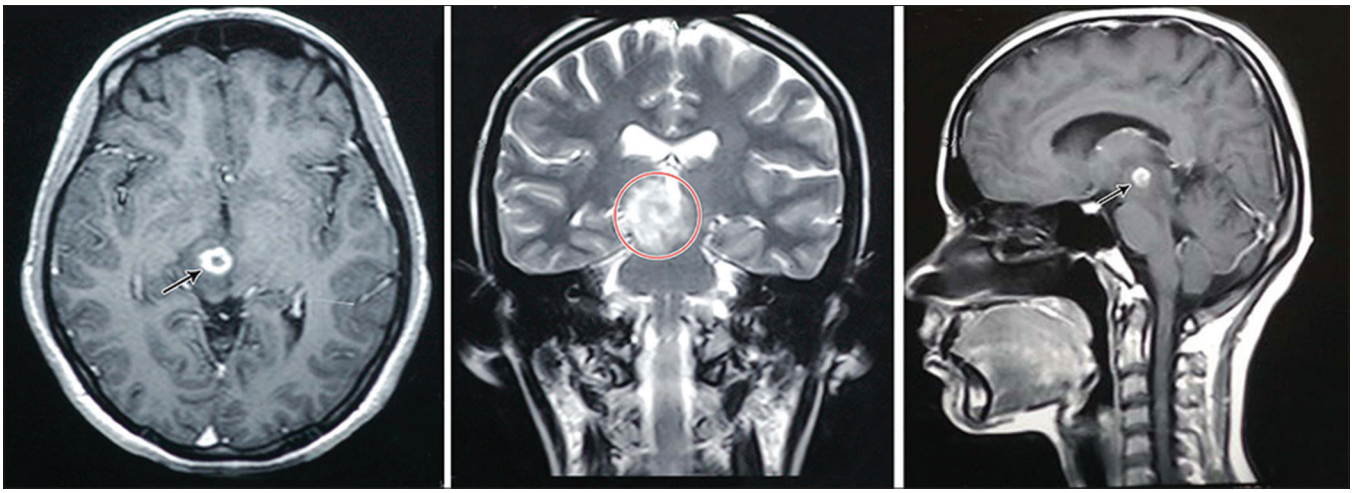

Figure 3: Magnetic resonance imaging brain with contrast suggestive of midbrain tuberculoma (1.4 cm × 1.2 cm) in right subthalamic region, crus cerebri, and periaqueductal area

isolated ring-enhancing hypointense lesion of 1.4 cm × 1.2 cm with a hyperintense rim on T2-weighted image, in the right subthalamic region, crus cerebri, and periaqueductal area, suggestive of midbrain tuberculoma [Fig. 3]. MR spectroscopy (MRS) showed high peak of lipids, increased choline, low N-acetyl aspartate (NAA), and creatinine with choline/creatinine ratio >1 which confirmed tuberculoma. Radiological search for tuberculous foci in other internal organs proved futile. Mantoux was positive confirming the exposure to tuberculous bacilli. She was started on low-dose

Figure 4: Response to therapy

steroids along with antitubercular therapy (ATT) AKT 4, which is a combination of ethambutol hydrochloride 800 mg, isoniazid 300 mg 2 tablets, pyrazinamide 750 mg 1 capsule, and rifampicin 450 mg. The patient was reviewed after 1 month. Diplopia had resolved and orthophoria was noted in primary position [Fig. 4]. Right pupil size and reaction had recovered by then. ATT was continued for 1½ years.[1] On regular follow-up, no side effects of ATT were noted.

Discussion

Tuberculomas are conglomerate, caseating foci that arise within the leptomeninges, ventricles, or the subdural space, though usually located within the substance of the brain. They develop during a period of hematogenous dissemination of *Mycobacterium tuberculosis* and commonly occur in patients who have a history of pulmonary tuberculosis, tuberculous meningitis, or both.[3,4] However, tuberculomas may also become symptomatic in patients with no history of tuberculosis and who have no evidence of pulmonary disease by chest X-ray.[5] In this latter group of patients, neurologic symptoms and signs may be the initial manifestation of underlying disseminated tuberculosis.[6]

The diagnosis of a tuberculoma is usually ascertained by pathology, neuroimaging, or clinical response to ATT.[7] MRI has revolutionized the imaging of tuberculomas and the diagnosis can be made with certainty to a reasonable extent. The lesions appear hypointense with or without central hyperintensity (due to caseous necrosis) or isointense that enhance after intravenous injection of contrast and are associated with significant surrounding brain edema.[8] MRS adds great value in the specific diagnosis of tuberculoma in case of ring-enhancing lesions, wherein it demonstrates a very high lipid peak, reduction in NAA, creatinine and a choline/creatinine ratio of >1. Lipid peak in MRS in a ring-enhancing lesion is very much specific for tuberculoma and is not found in any case of neurocysticercosis, the other common differential diagnosis of a ring-enhancing lesion, which also demonstrates intralesional scolex in imaging studies.[9]

Our patient had the typical MRI findings of tuberculoma, further confirmed by MRS. The clinical picture in our case favors the proposed transverse neuroanatomic organization of the fascicular fibers of the oculomotor nerve (mediolateral somatotopy), with superior rectus and inferior rectus being the most lateral and caudal, and the pupilloconstrictor fibers and the inferior rectus being the most medial and rostral. Thus, a lesion located in this area caused isolated inferior rectus palsy.

Nystagmus is thought to be due to imbalance of the vertical vestibulo-ocular reflex and otoliths. Close proximity of the lesion to rostral interstitial nucleus of the MLF and the interstitial nucleus of Cajal may have caused nystagmus in this case.[10]

Saxena *et al*. had described a case of supranuclear gaze palsy without diplopia as a manifestation of a tuberculous brain stem lesion.[11] In another report by Monteiro and Coppeto, the patient presented with diplopia and abnormal eye movements and had a supranuclear gaze palsy together with reduced abduction of the eye due to tuberculomas of brain stem and cerebellum.[12] Furthermore, one-and-a-half syndrome has been reported due to pons and midbrain tuberculomas.[13]

Conclusion

To the best of our knowledge, this is the first case to be reported with pupil-involving third nerve palsy along with downbeat nystagmus in a case of brain-stem tuberculoma. Most of the patients respond well with medical therapy and surgical intervention is not required. Role of steroids is controversial.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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