Weber syndrome secondary to brain stem tuberculoma

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This case report describes a rare presentation of presumed brain stem tuberculoma in a 28-year-old male who presented with acute onset of third cranial nerve palsy with contralateral hemiparesis (Weber syndrome) and upgaze palsy. Isolated midbrain tuberculoma is rare, presenting with varied clinical manifestations and radiological findings posing as a diagnostic dilemma. Weber syndrome is commonly caused by midbrain infarct secondary to occlusion of branches of the posterior cerebral artery and rarely from a tuberculoma. The patient is a case of disseminated tuberculosis with granuloma in midbrain causing pressure effect, thereby presenting with features consistent with Weber syndrome and upgaze palsy. The patient had good recovery with antitubercular treatment and systemic steroids.

Key words: Brain stem tuberculoma, third cranial nerve palsy, Weber syndrome

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Tuberculosis (TB) of the central nervous system (CNS) accounts for 20%–30% of intracranial space-occupying lesions (ISOL) in developing countries.[1] Tuberculoma constitutes 1.4% of the incidence of CNS TB and occurs secondary to hematogenous dissemination from elsewhere in the body.[2] Isolated brain stem tuberculoma is rare and constitutes 5% of all intracranial tuberculomas in endemic

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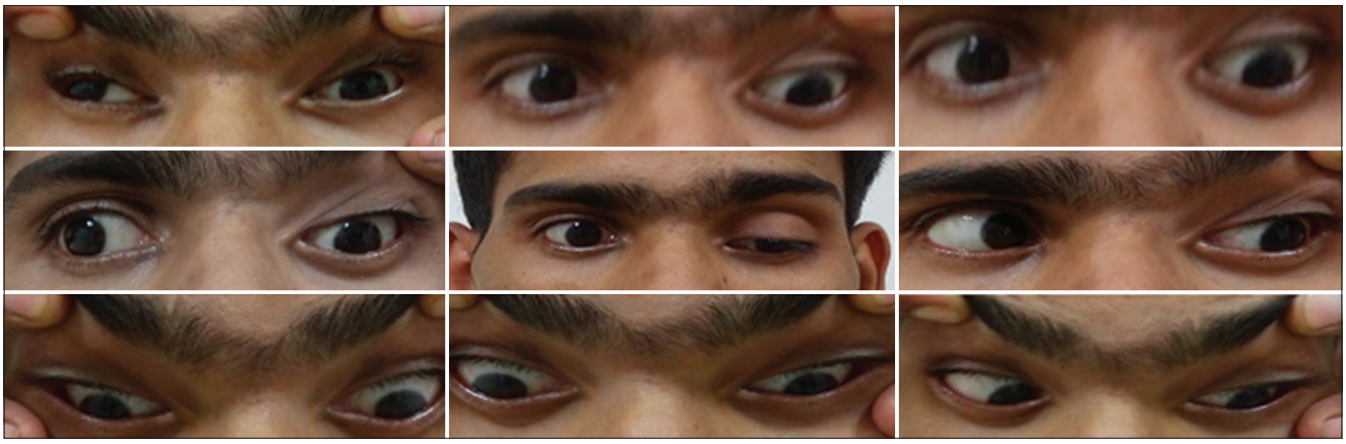

Figure 1: Extraocular movements in nine gazes with limitation of upgaze in both the eyes along with left eye movements limited on adduction and depression

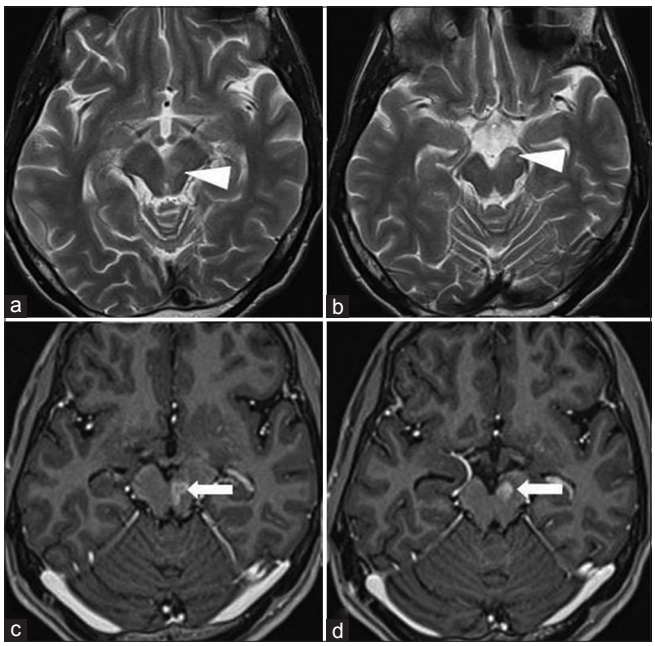

Figure 2: Cranial magnetic resonance imaging axial T2-weighted image shows hyperintensity in the midbrain on the left side (white arrowhead in a and b). Postcontrast axial T1-weighted image (c and d) shows nodular and peripheral enhancing conglomerate lesions (white arrows) in the ventromedial aspect midbrain and left cerebral peduncle

areas.[3,4] Clinical manifestations can be varied depending largely on their location and number.[5-8] Patients can present with signs of raised intracranial pressure or focal neurological deficits such as cranial nerve palsy and motor and sensory symptoms. The radiological features are also nonspecific leading to misdiagnosis in few cases.

Any midbrain lesion causing a combination of ipsilateral third nerve palsy and contralateral hemiplegia is known as Weber syndrome which was first described and named after Weber.[9] It is commonly caused by vascular pathology such as an infarction or aneurysm, tumor, and demyelination.[10] Weber syndrome secondary to brain stem tuberculoma is rarely reported[11] highlighting the diagnostic challenge of this case.

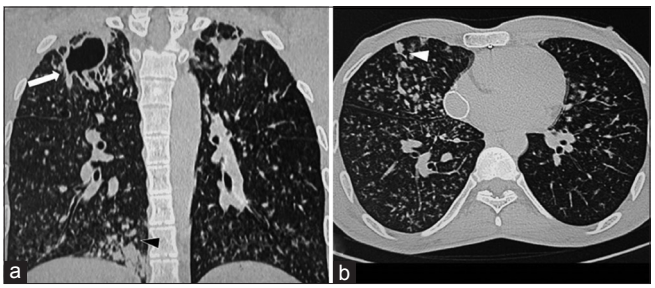

Figure 3: Coronal reformatted (a) and axial (b) image of lungs shows fibrocavitary lesion in the right upper lobe (white arrow in a) and nodular lesion in the left upper lobe. Multiple small nodules are seen diffusely in both the lungs (arrowheads in a and b), some showing tree-in-bud appearance (white arrowhead in b)

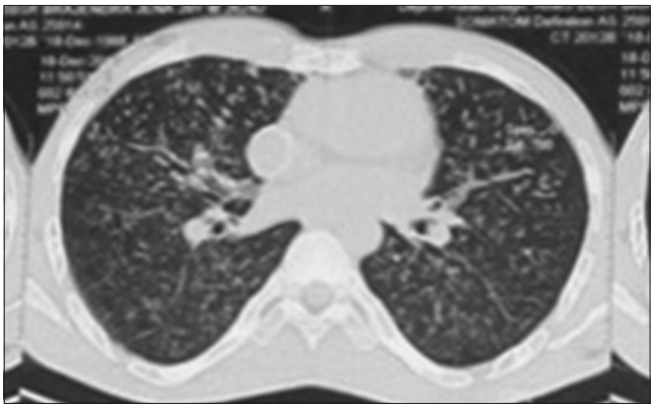

Figure 4: High-resolution computed tomography thorax shows fibrocavitary lesion with traction bronchiectasis with areas of consolidation suggestive of old pulmonary tuberculosis with reactivation

Case Report

A 28-year-old male presented to emergency department with sudden onset of headache, tingling sensation in the right upper and lower limb, and drooping of the left eyelid for 2 days. There was a history of intermittent fever, cough, and weight loss for the past 15 days. Medical history revealed congenital heart disease (Fallot physiology) which was operated at the age of 14 years.

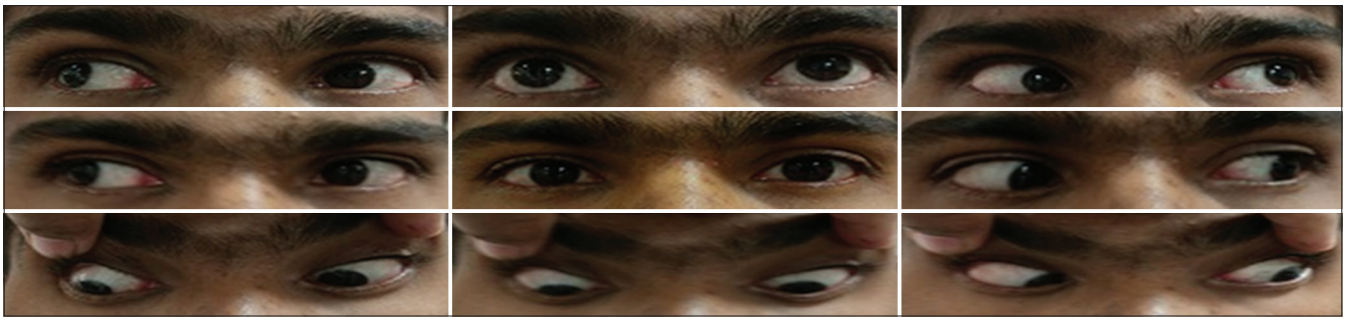

Figure 5: Posttreatment extraocular movements full in all gazes in both the eyes except for limitation of upgaze in both the eyes

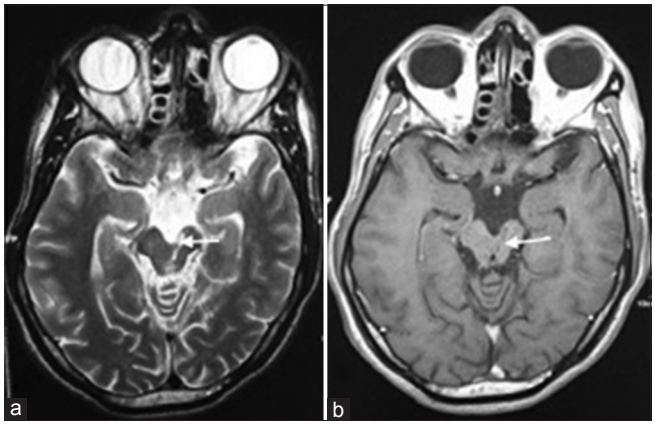

Figure 6: Posttreatment magnetic resonance imaging brain shows no enhancement of the lesion on contrast (white arrow in a and b)

The patient was conscious and oriented. His best-corrected visual acuity was 6/6 P in both the eyes. Ocular examination of the right eye was normal except for limitation of elevation in upgaze. The left eye showed complete ptosis with exotropia and hypotropia with limitation of adduction, elevation, and depression [Fig. 1]. Convergence was impaired. Right eye pupil was 3 mm, reacting while left eye pupil was 4 mm with sluggish reaction and anisocoria increasing in bright light. Slit-lamp biomicroscopy of the anterior segment and fundus was normal. Color vision was normal in both the eyes. The patient could not complete the visual field testing and diplopia charting.

Neurological examination revealed right hemiparesis with hemiplegic gait and reduced touch sensation. Reflexes were brisk on the right side with extensor plantar response. Coordination was normal, and no other cranial nerve abnormalities detected. Examinations of pulmonary and cardiovascular systems were within normal limits.

A provisional diagnosis of cardioembolic brain stem stroke secondary to cardiac disease presenting as Weber syndrome and upgaze palsy was made and started on antiplatelet medication.

However, after diagnostic investigations including magnetic resonance imaging (MRI) of the brain and high-resolution computed tomography thorax, the diagnosis was revised for the following two reasons. (1) MRI brain T2 and fluid-attenuated inversion recovery showed hyperintense lesions in the left ventromedial midbrain and ipsilateral cerebral peduncle [Fig. 2]. Peripheral and nodular enhancement was noted in postcontrast T1-weighted image. Conglomerate nodular and peripherally enhancing lesions favor the diagnosis of tuberculoma. There was minimal perilesional edema. (2) Noncontrast CT scan of the lungs showed fibrocavitary lesion in the right upper lobe with traction bronchiectasis and nodular lesion in the left upper lobe. Multiple small nodules were seen diffusely in bilateral lung fields and some having tree-in-bud appearance [Figs. 3 and 4]. Hence, a diagnosis of reactivation of pulmonary TB was made. To further confirm the diagnosis, magnetic resonance spectroscopy (MRS) was advised, but the patient could not afford it.

The clinical examination showed pupil involving left third nerve palsy with upgaze palsy together with right hemiparesis which favored the final diagnosis of Weber syndrome with upgaze palsy. The midbrain lesion was presumed to be of tubercular etiology. Treatment was started with antitubercular drugs comprising of isoniazid 75 mg, rifampicin 150 mg, ethambutol 275 mg, and pyrazinamide 400 mg along with intravenous streptomycin 0.75 g and systemic steroids. After 6 months of treatment, the patient had complete recovery of movements in all gazes except for upgaze in both the eyes [Fig. 5]. Posttreatment MRI scan showed resolution of the enhancing mass [Fig. 6].

Discussion

Intracranial tuberculoma is usually located in the highly vascular areas of the brain such as the cerebral and cerebellar hemisphere and rarely in the brain stem.[10,11] Isolated brain stem tuberculoma presents with varied clinical presentations such as one-and-a-half syndrome, horizontal gaze palsy, and bilateral internal ophthalmoplegia.[10] The critical location of the lesion in the left ventromedial aspect of the midbrain with perilesional edema in the patient explains the clinical manifestations of the third cranial nerve palsy with contralateral hemiparesis, thus presenting as Weber syndrome.

Talamás *et al.*[2] reported 11 cases of brain stem tuberculoma, of which three cases presented with features consistent with Weber syndrome along with neurological deficits. All the three patients had midbrain granuloma on neuroimaging, and none had any evidence of TB outside CNS. Granuloma disappeared following treatment in all the three patients; however, clinical symptoms improved in only two cases.

Rajshekhar and Chandy[4] reported six cases of isolated brain stem tuberculoma, and among them, only one patient had a midbrain lesion along with hydrocephalus on imaging. Patients presented with features of pyramidal tract, third and

seventh cranial nerve involvement. No evidence of pulmonary TB was seen on chest radiography. Stereotactic biopsy along with ventriculoperitoneal shunt was done. Patients had symptomatic improvement with antitubercular therapy.

Khalil *et al*.[11] reported about the association of Weber syndrome with vertical gaze palsy. He explained it by the fact that vertical eye movements are generated in the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) which also lies in the midbrain, dorsal to the red nucleus. The clinical presentation of the patient also supports the accepted topographical fascicular arrangement of the oculomotor nerve and riMLF in the midbrain.

Both the diagnosis and management of this case were challenging. The initial clinical presentation was concerning for stroke as well as any ISOL. However, imaging showed features of reactivation of TB in the chest and a ring-enhancing lesion in the midbrain favoring infectious etiology over a vascular pathology such as infarction or thromboembolic phenomenon. MRS plays a specific role in the diagnosis of tuberculoma as it shows lipid deposits and differentiates it from brain abscess, neurocysticercosis, metastasis, and demyelinating diseases which could have aided in our diagnosis.[10]

This case could be a reactivation of TB, but reinfection is a possibility. The Centers for Disease Control and Prevention recommends 12 months of treatment for CNS TB.[12] However, it can be tailored as per the response and radiological findings. Systemic corticosteroids can be indicated as adjuvant therapy when there is perilesional edema or paradoxical progression during treatment.[7]

Conclusion

To conclude, brain stem tuberculoma is a rare cause of Weber syndrome and difficult to diagnose without tissue confirmation. A high index of suspicion is warranted in endemic areas. The patient is one of those rare cases of CNS tuberculoma presenting as a syndrome associated with cranial nerve palsy. Ophthalmologists must be aware of this potentially curable disease which requires integration of clinical and radiological diagnosis and timely management.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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