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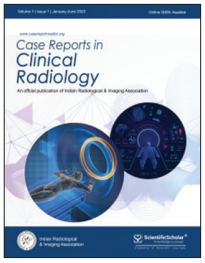

Case Series

Two rare central nervous system manifestations of a common disease

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Received: 05 February 2024
Accepted: 04 April 2024
EPub Ahead of Print: 08 June 2024
Published: 13 July 2024

DOI
10.25259/CRCR_15_2024

Quick Response Code:

ABSTRACT

Tuberculosis (TB) is an endemic infection caused by *Mycobacterium tuberculosis*. Various manifestations of the central nervous system (CNS) TB include cerebritis, cerebral abscess, meningitis, tuberculoma, and encephalopathy. Isolated intraventricular tuberculoma and hypertrophic pachymeningitis are two rare presentations of CNS TB infections. Intraventricular tuberculoma is rare, of which third ventricular location is extremely rare and only two cases of third ventricular tuberculoma are reported in the literature. Although leptomeningitis is common, isolated hypertrophic pachymeningitis is an uncommon presentation of CNS TB.

Keywords: Third ventricular tuberculoma, Central nervous system tuberculosis, Intraventricular tuberculoma, Hypertrophic pachymeningitis

INTRODUCTION

Tuberculosis (TB) is an endemic infection in India which is caused by *Mycobacterium tuberculosis* (Mtb). As per the global TB report of 2017, the incidence of TB in India was approximately 2–3 million, and central nervous system (CNS) TB accounts for 1–10% of all TB cases.[1-3] Herein, we report two rare CNS presentations of this common disease, which are isolated intraventricular tuberculoma and hypertrophic pachymeningitis.[4,5]

CASE SERIES

Case 1

A 27-year-old female patient presented to our emergency department with complaints of fever, headache, giddiness, and vomiting for the past 1 week. There was no significant past medical or surgical history. On general examination, the patient was conscious, oriented, and febrile, and Glasgow Coma Scale (GCS)-15/15 and vitals were within normal limits. On physical examination, neck rigidity was present, and Kernig's and Brudzinski's signs were positive. Systemic examination was normal.

Non-contrast computed tomography (CT) brain axial image at the level of the roof of the third ventricle showed a relatively well-defined hyper-isodense lesion with no calcifications or obstructive hydrocephalus [Figure 1]. Contrast-enhanced CT brain axial image showed intense homogeneous enhancement of the lesion [Figure 1]. Magnetic resonance imaging (MRI) brain with contrast was suggested for further evaluation.

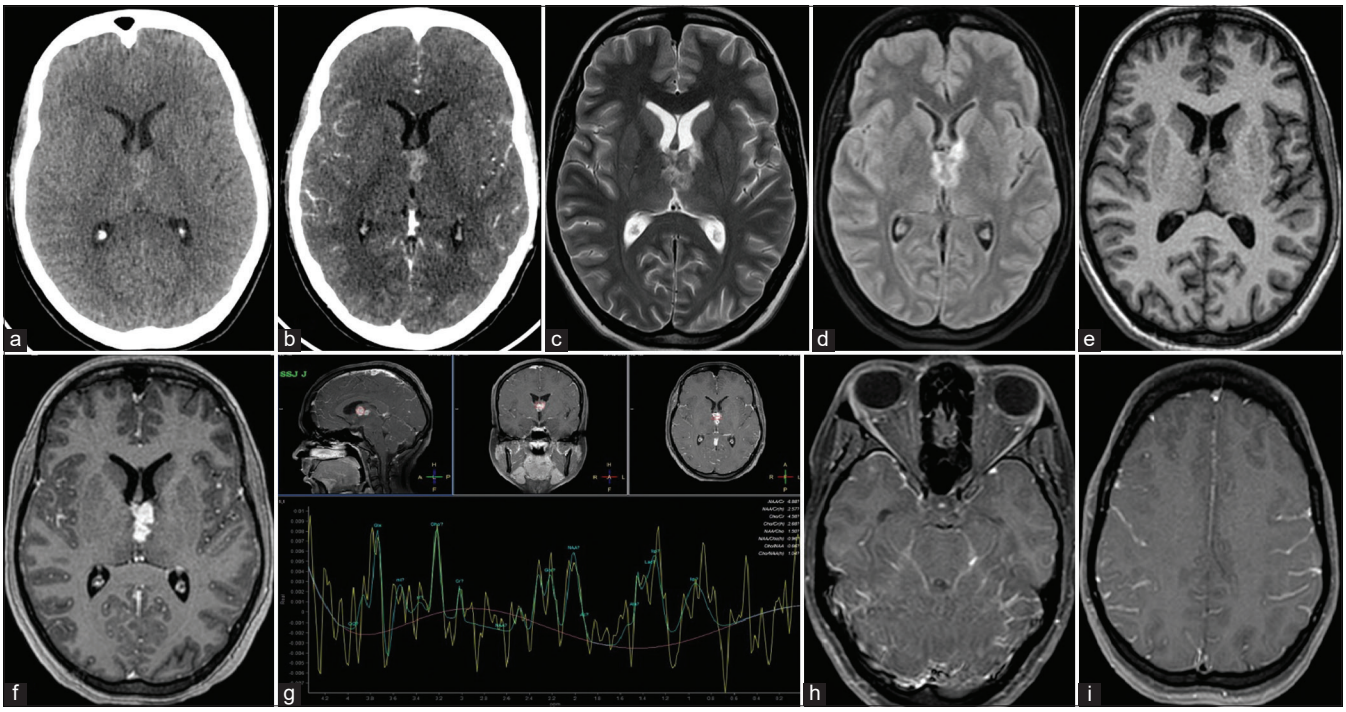

Figure 1: Computed tomography brain plain (a) and contrast (b) axial sections at the level of roof of the third ventricle show a relatively well-defined iso-hyperdense lesion with intense homogeneous post-contrast enhancement. Magnetic resonance imaging (MRI) brain T2 (c) and FLAIR (d) axial sections at the level of roof of the third ventricle show a T2 hypointense lesion which does not show suppression on fluid-attenuated inversion recovery (FLAIR) image. MRI brain T1 (e) and T1 gadolinium contrast (f) axial sections at the level of roof of the third ventricle show an iso-intense lesion which shows intense homogeneous post-contrast enhancement. Magnetic resonance-spectroscopy (g) shows a lipid lactate peak at 1.3 ppm. MRI brain T1 contrast axial images at the level of basal cisterns (h) and fronto-parietal sulci (i) show enhancement of basal cisterns and lepto-meningeal thickening/enhancement.

T2 axial image at the level of the roof of third ventricle showed a hypo-intense lesion which on fluid-attenuated inversion recovery (FLAIR) axial image showed no suppression [Figure 1]. T1 axial image at the level of the roof of third ventricle showed an iso-intense lesion which on the T1 contrast axial image showed intense homogeneous contrast enhancement [Figure 1]. Magnetic resonance spectroscopy (MRS) image showed a lipid lactate peak at 1.3 ppm [Figure 1]. T1 contrast axial images at the level of basal cisterns and fronto-parietal sulci showed enhancement of basal cisterns and lepto-meningeal thickening/enhancement [Figure 1]. Based on the above imaging finding, possibility of intraventricular tuberculoma was considered. Cerebrospinal fluid (CSF) analysis was done which showed increased protein (362 mg/dL) and raised adenosine deaminase (ADA) values (9.5 IU/L) consistent with TB infection.

Due to the smaller size of lesion and the absence of hydrocephalus, a course of anti-tuberculosis treatment (ATT) was administered and the patient came for review after 3 months. Follow-up CT brain contrast axial image showed a decrease in size and intensity of enhancement of the lesion [Figure 2]. Surgical intervention was not done as the lesion was showing a response to ATT and no signs of obstructive hydrocephalus were present. Follow-up CT brain with contrast was done after 6 months of ATT which showed a further decrease in size and intensity of enhancement of the intraventricular lesion [Figure 2]. The patient is on serial follow-up.

Case 2

A 54-year-old male patient came with complaints of left-sided headaches for 4 months on and off which aggravated for the past 1 month. He also had complaints of decreased appetite and nausea for 2 days. There is no evidence of trauma, fever, or seizures. There is no clinical evidence of malignancy. On examination, the patient was conscious, oriented, and GCS – 15/15. There are no neck rigidity and no nystagmus and power: Normal. Systemic examination was normal. CSF study was done, except for mildly elevated lymphocytes and sugar levels, otherwise normal. Total blood count showed mildly elevated white blood cells.

The patient was referred for imaging modalities. Plain and contrast-enhanced CT brain showed white matter edema in the left temporal lobe with enhancement and thickening of the left tentorium cerebelli [Figure 3]. There is no evidence of enhancing mass lesions or dural venous sinus thrombosis [Figure 3]. There is no evidence of bony erosions

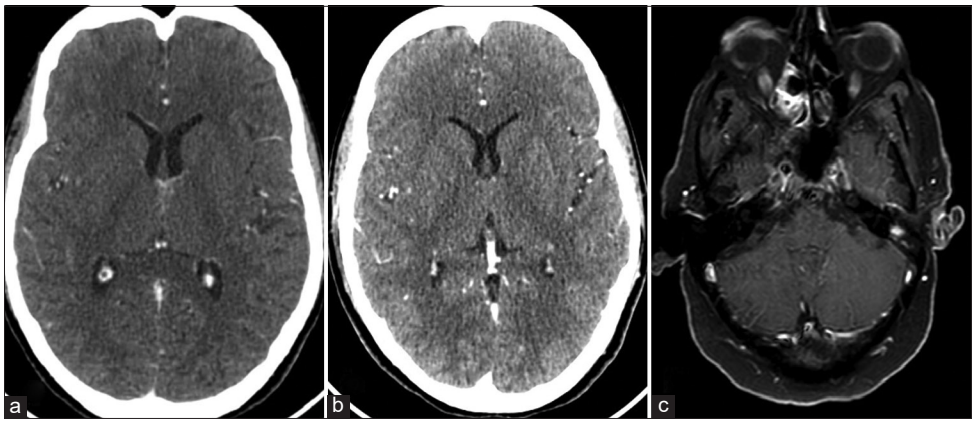

Figure 2: (a) Follow-up computed tomography (CT) brain contrast axial image of case 1 after 3 months of ATT at the level of the roof of the third ventricle shows a reduction in size and intensity of enhancement of the lesion. (b) Follow-up CT brain contrasts the axial image of case 1 after 6 months of ATT at the level of the roof of the third ventricle shows further reduction in size and intensity of enhancement of the lesion. (c) Follow-up magnetic resonance imaging brain contrasts the axial image of case 2 after 3 months of ATT shows a significant reduction in thickening and enhancement of pachymeninges in the left temporal region. ATT: anti-tuberculosis treatment.

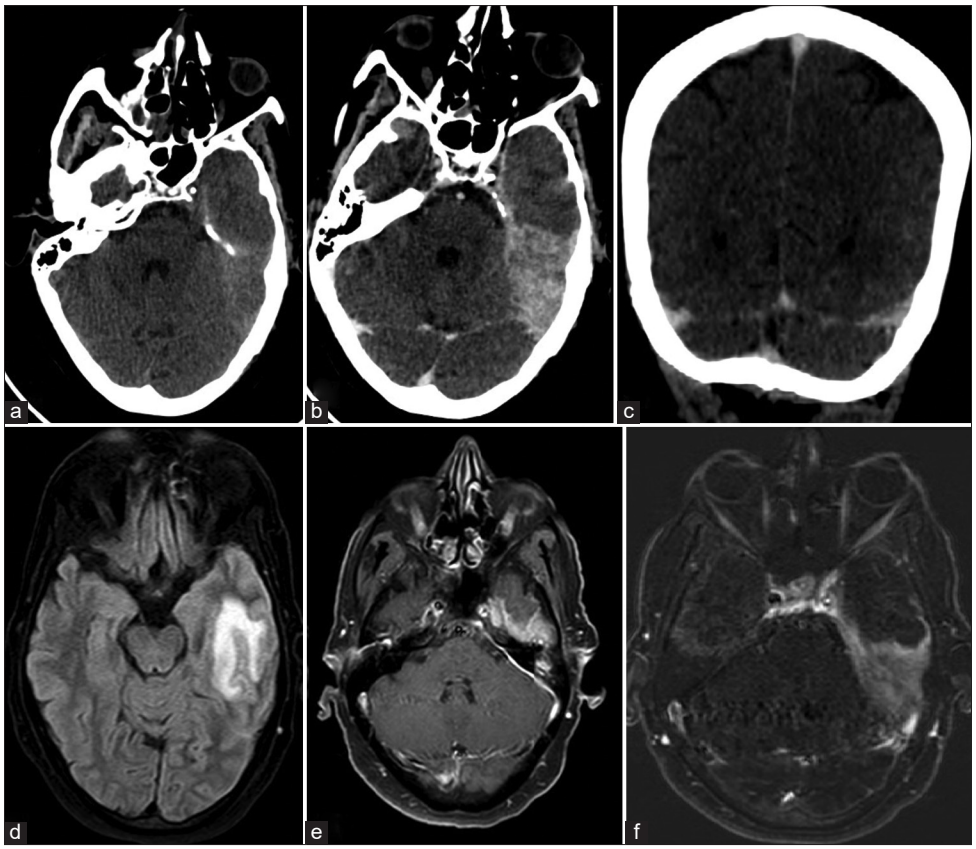

Figure 3: (a) Computed tomography (CT) brain plain and (b) contrast axial sections show thick hyperdense left tentorium cerebelli with post-contrast enhancement and minimal left temporal lobe edema. (c) CT contrast coronal section shows no evidence of enhancing mass lesions or dural venous sinus thrombosis. (d) Magnetic resonance imaging (MRI) of brain flair (e) axial section shows hyperintensity in the left temporal lobe. MRI brain T1 gadolinium contrast and (f) T1 post-contrast subtracted image axial section shows focal abnormal pachymeningeal thickening and enhancement in the left temporal region.

or skull base osteomyelitis. MRI was suggested to rule out meningoencephalitis.

Areas of FLAIR hyperintensities noted in left temporal lobe without evidence of diffusion restriction [Figure 3]. Post-contrast study shows focal abnormal pachymeningeal thickening and enhancement in the left temporal region [Figure 3]. The patient was diagnosed with hypertrophic pachymeningitis with temporal lobe edema. The screening panel for auto-immune disease was negative. The patient was empirically started ATT and follow-up imaging after 3 months showed a significant reduction in thickening and enhancement of pachymeninges in the left temporal region [Figure 2].

DISCUSSION

TB is an endemic infection in India which is caused by Mtb. The lung is the predominant site of involvement in tubercular infection and extra-pulmonary site involvement occurs in 20–45% of cases.[6] Risk factors for extra-pulmonary TB include HIV infection, diabetes mellitus, low socioeconomic status, and other comorbidities.[7] The extra-pulmonary sites include the lymph nodes, abdomen, CNS, genitourinary system, bones, and joints.[6,7] CNS TB accounts for 1–10% of all TB cases.[1]

Various manifestations of CNS TB include cerebritis, cerebral abscess, meningitis, tuberculoma, and encephalopathy.[6] CNS tuberculoma is one of the serious complications occurring following CNS TB infections. The most common location of intracranial tuberculoma is cerebral parenchyma.[2,3] Other locations of tuberculoma include sellar and suprasellar regions,[8] brainstem,[9] cerebellum,[10] and intraventricular.[11]

Intraventricular tuberculoma is rare,[4,5] of which third ventricular location is extremely rare.[4,5] The blood circulation of the ventricular system acts as an effective barrier to the spread of infections. The most common route of spread of tubercle bacilli to ventricles is assumed to be hematogenous spread through the choroid plexus.[12] CSF analysis is key for the diagnosis of intracranial TB but it is not much useful in the diagnosis of intracranial tuberculoma. Imaging plays an important role in the diagnosis and to avoid unnecessary surgical intervention.

Intracranial tuberculoma is typical of granulomatous etiology which is initially non-caseating granuloma and eventually progresses to caseating granuloma with central liquefaction. There are four stages of tuberculoma: Non-caseating granuloma, caseating granuloma, caseating granuloma with central liquefaction, and calcified granuloma. MRI with gadolinium contrast is the choice of investigation in the diagnosis of tuberculoma and is superior to CT.[13] The imaging features of tuberculoma vary depending on the stage of presentation. On non-contrast CT, tuberculoma may appear as an isodense, hypodense, or even as a hyperdense area, which on post-contrast study shows homogeneous or ring enhancement.[14] Non-caseating granuloma appears iso-hypointense on T1, hyperintense on T2, with homogeneous enhancement on T1 gadolinium contrast and shows no diffusion restriction. Caseating granuloma and caseating granuloma with central liquefaction show specific T1 hyperintense rim, however, caseating granuloma with central liquefaction shows central hyperintensity on T2 with ring enhancement on T1 gadolinium contrast and may or may not show diffusion restriction. Calcified granuloma appears hypointense on T2 and shows no perilesional edema or contrast enhancement.[13] On MRS, lactate peak at 1.3 ppm is characteristic of tuberculoma and in recent studies, guanidinoacetate peak at 3.8 ppm is also found to be characteristic of tuberculoma.[15]

The differentials for enhancing intraventricular lesions include ependymoma, subependymoma, intraventricular meningioma, subependymal giant cell astrocytoma (SEGA), neurocysticercosis, and metastases. Ependymoma occurs commonly in children and young adults with a floor of the fourth ventricle being the most common site. They show heterogeneous post-contrast enhancement with choline peak on MRS. Subependymoma occurs in middle-aged adults and the fourth ventricle is their most common site. They usually do not show enhancement or at times may show mild enhancement.

Intraventricular meningioma occurs in middle-to-old aged adults and lateral ventricles are their most common site. They show vivid post-contrast enhancement with an alanine peak on MRS. SEGA occurs in patients with tuberous sclerosis and the foramen of monro is their classical location. They show heterogeneous post-contrast enhancement. Neurocysticercosis occurs mostly in sulcal and subarachnoid spaces. They appear hyperintense on T2 with eccentric T2 hypointense scolex and ring enhancement on post-contrast study. On MRS, they show a succinate peak at 2.4 ppm. Metastases may show ring enhancement on post-contrast study mimicking caseating granuloma with central liquefaction, however, a prominent choline peak is noted in the former [Table 1].

Hypertrophic pachymeningitis is a rare form of chronic fibrosing inflammatory disease of cranial or spinal dura matter. It may be either focal or diffuse. It may be primary where etiology is unknown or secondary to conditions such as infections (TB, syphilis), mastoiditis, and inflammatory conditions such as rheumatoid arthritis, sarcoidosis, Wegener's granulomatosis, post-radiation, and autoimmune disorders. Usually, patients present with headache, symptoms related to cranial nerve involvement such as visual disturbances, loss of vision, hard of hearing, hearing loss, and seizures.[16,17]

Miyamoto *et al*. and Suárez-Calvet *et al*. have reported two rare cases of tuberculous hypertrophic pachymeningitis which was confirmed by cervical lymph node biopsy. The most likely route of dissemination is lymphatic spread as there are abundant lymphatic vessels in the dural sheath.[18,19] Imaging

plays an important role in the diagnosis of hypertrophic pachymeningitis. Unlike diffuse leptomeningeal thickening or basal exudates which are commonly seen in CNS TB, this condition is noted by focal thickening of pachymeninges which shows post-contrast enhancement. The common differential diagnosis includes meningioma en plaque which is characterized by bony changes. Isolated pachymeningitis in TB is extremely rare and diagnosis of this condition is important to avoid unnecessary surgical interventions.[20,21]

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for intraventricular lesions are described in Table 1.

Table 1: The differential diagnosis for intraventricular lesions.

Differentials for intraventricular lesions	Differentiating features
1. Ependymoma	• Common in children and young adults with the floor of the fourth ventricle being the most common site. • They show heterogeneous post-contrast enhancement with choline peak on MRS.
2. Subependymoma	• They occur in middle-aged adults and the fourth ventricle is their most common site. • They usually do not show enhancement or at times may show mild enhancement.
3. Intraventricular meningioma	• They occur in middle-to-old aged adults and lateral ventricles are their most common site. • They show vivid post-contrast enhancement with an alanine peak on MRS.
4. Subependymal giant cell astrocytoma	• They occur in patients with tuberous sclerosis and the foramen of monro is their classical location. • They show heterogeneous post-contrast enhancement.
5. Neurocysticercosis	• They occur mostly in sulcal and subarachnoid spaces. • They appear hyperintense on T2 with eccentric T2 hypointense scolex and ring enhancement on post-contrast study. • On MRS they show a succinate peak at 2.4 ppm.
6. Metastases	• Heterogeneous post-contrast enhancement or at times shows ring enhancement. • Choline peak on MRS.

MRS: Magnetic resonance spectroscopy

CONCLUSION

To conclude, isolated intraventricular tuberculoma and hypertrophic pachymeningitis are two uncommon presentations of TB which can be of diagnostic dilemma. It is important to diagnose these conditions as they can be treated with ATT without surgical interventions.

TEACHING POINTS

1. Isolated intraventricular tuberculoma is a rare and diagnostic challenge which has to be kept as a differential for intraventricular lesions under appropriate clinical scenario
2. Hypertrophic pachymeningitis is a rare presentation of CNS TB, which needs to be differentiated from other etiologies.

MCQs

1. All are differentials for intraventricular enhancing lesions except
   a. Ependymoma
   b. Intraventricular meningioma
   c. Tuberculoma
   d. SEGA
   e. None of the above

Answer Key: e

2. Which of the peak is characteristic of tuberculoma on MRS?
   a. Lipid lactate peak
   b. Alanine peak
   c. Succinate peak
   d. None of the above

Answer Key: a

3. What are the causes of hypertrophic pachymeningitis?
   a. Idiopathic
   b. TB
   c. Autoimmune disorders
   d. All of the above

Answer Key: d

Acknowledgment

We acknowledge Dr. N. Arun Kumar MCh, consultant neurosurgeon and Dr. N. Arathi MD pathology , Meenakshi hospital Thanjavur for their valuable contribution towards clinical and pathological findings.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Shanmuga Jayanthan S, Nadanasadharam K, Rupesh G, Sumathi MS. Two rare central nervous system manifestations of a common disease. Case Rep Clin Radiol. 2024;2:81-6. doi: 10.25259/CRCR_15_2024