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Case Report

Focal dystonia and ataxic hemiparesis as the initial presentation of a thalamic tuberculoma: A diagnostic challenge in an immunocompetent pediatric patient

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ABSTRACT

Background: Central nervous system (CNS) tuberculomas are rare and account for approximately 1% of all tuberculosis (TB) cases. These intracranial lesions are more commonly observed in immunocompromised individuals, often as part of disseminated miliary TB or after latent infection reactivation. This case report presents the occurrence of a thalamic tuberculoma in an immunocompetent girl.

Case Description: An 11-year-old girl presented with a 3-month history of progressive right-sided ataxic hemiparesis, hand dystonia/thalamic hand, and headache. There was only a mildly elevated erythrocyte sedimentation rate (25 mm/h.), and her remaining biochemistry and vitals were unremarkable. Magnetic resonance imaging (MRI) brain revealed an ill-defined intra-axial heterogeneous lobulated lesion with crenated margins involving the thalamus and the posterior limb of the internal capsule with significant vasogenic edema. Given the clinical picture, the working diagnosis was a high-grade brain tumor. Due to the absence of a viable operative corridor for a meaningful resection and the diagnostic uncertainty, a stereotactic biopsy was performed, and histopathological analysis confirmed the presence of granulomas consistent with TB. A human immunodeficiency virus test (negative) and interferon-gamma release assay (positive) were then obtained. The patient was commenced on a regimen of anti-TB drugs with a tapering steroid dose. At 8 months, her most recent MRI showed a significant reduction in the size of her tuberculoma, and there is a complete resolution of her hand dystonia and hemiparesis to allow for independence in her activities of daily living.

Conclusion: This report emphasizes the importance of considering causes other than degenerative, vascular, or neoplasms in patients with hemiparesis with dystonia. CNS tuberculomas can present as such without prior history or specific clinical symptoms of TB, making them a diagnostic challenge. In cases with such uncertainty regarding the nature of an intracranial lesion and the role of resection, a stereotactic biopsy is invaluable.

Keywords: Ataxic hemiparesis, Immune competent, Intracranial tuberculoma, Lacunary syndrome, Thalamic hand, Thalamus

INTRODUCTION

Neurological deficits from ischemia, infarction, and mass lesions are common presentations in neurology/neurosurgery. Lacunar syndrome is a neurological deficit from a deep brain lesion, most commonly ischemia/infarctions.[6] Ataxic hemiparesis is a manifestation of lacunar syndrome presenting as unilateral pyramidal weakness and ipsilateral ataxia and can occur from lesions to several cerebral structures. The thalamus and internal capsule are two well-known locations where vascular lesions can result in dystonia and hemiataxia as a common presentation.[3] In pediatric patients, however, cerebrovascular disease is less common a cause for ataxic hemiparesis and focal dystonia than neurodegenerative diseases and space-occupying lesions such as brain tumors. Central nervous system (CNS) tuberculomas are rare intracranial lesions, especially in immunocompetent Bacillus Calmette-Guérin (BCG)-vaccinated individuals.[5] Intracranial tuberculomas are most commonly intra-axial/intraparenchymal and are likely to be preceded by a history of tuberculosis (TB), clinically as part of reactivated TB or miliary TB, when the disease is disseminated in multiple organs.[5,7] The authors report an 11 years old, BCG vaccinated, immunocompetent healthy girl, who, *de novo*, presented with a 3-month history of right-sided ataxic hemiparesis and dystonia, which was eventually found to be a result of an isolated thalamic tuberculoma.

CASE DESCRIPTION

An 11-year-old female presented to our outpatient department complaining of right-sided weakness and headache, which had progressively worsened over 2–3 months. A thorough clinical examination revealed power 4/5 (Medical Research Council scale) in her right limbs with an upgoing plantar. The patient had an ataxic gait and focal dystonia in the form of a "thalamic hand" [Figure 1a (preoperative), Figures 1b and 1c showing complete resolution at last follow-up]. A comprehensive history revealed no relevant past medical, pharmacological, or family history. Routine investigations, including blood biochemistry, only revealed a mildly elevated erythrocyte sedimentation rate (ESR, 25 mm/h.). A non-contrast computed tomography head was performed the same day, revealing a hypodense mass in the left thalamic region, and the patient was admitted to our neurosurgery department. The following day, a magnetic resonance imaging (MRI) brain, revealed an ill-defined intra-axial heterogeneous lobulated lesion with crenated margins involving the right thalamus, the posterior limb of the internal capsule ipsilaterally, and several other neural structures that were being compressed from vasogenic edema causing mass-effect [Figure 2]. The patient was, thus, commenced on dexamethasone. Figure 2 shows her baseline MRI and its associated findings.

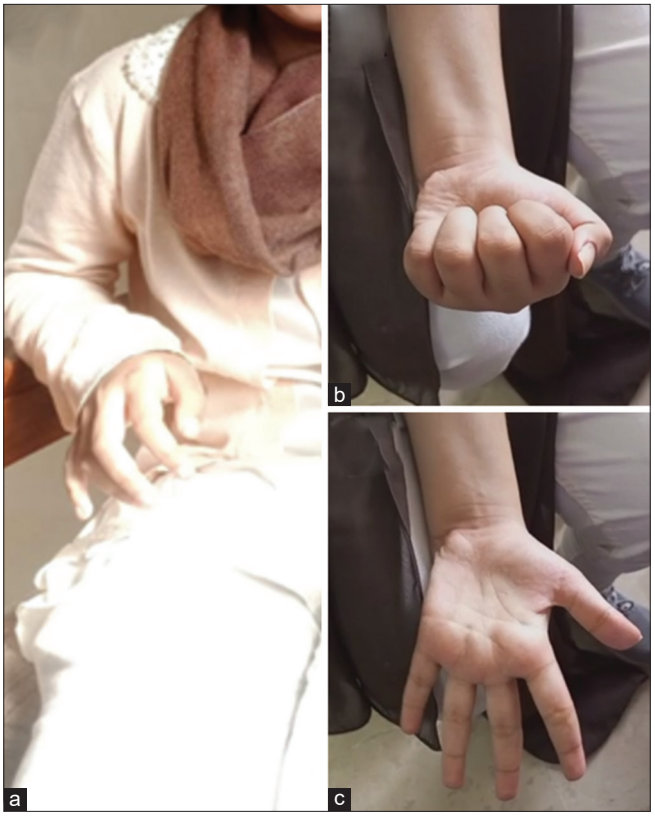

Figure 1: Left column (a) showing the patient's preoperative dystonic, "thalamic," hand that was contracted in this position indefinitely from November 2022. The right column (b and c) from June 2023 illustrates the complete resolution of the dystonia and a full range of movements and power of the initially hemiparetic side.

Considering the patient's clinical history and young age, the radiologist reported the differentials as a supratentorial primitive neuroectodermal tumor, extraventricular neurocytoma, or any other high-grade glioma. We, initially, planned to excise the lesion, but given the deep-seated nature of the lesion and the lack of a viable operative corridor, it was decided to perform a stereotactic biopsy as the neurosurgical working differential was of a high-grade tumor; we did not want to risk neurological deficits in the face of our provisional diagnosis and planned to utilize chemo/radiotherapy as/if indicated. A stereotactic biopsy was performed on the 4th day using an inomed frame. Biopsy microspecimens revealed caseous necrosis and necrotizing granulomatous inflammation. Histologically, scattered granulomas comprised epithelioid histiocytes with admixed lymphocytes, plasma cells, and giant cells were observed. Immunohistochemical staining was positive for CD3 (indicating reactivated T-cells), CD20 (indicating reactivated B cells), negative for Grocott methenamine silver stain (fungus), and negative for Ziehl-Neelsen stain. As the patient belonged to an impoverished socioeconomic background, TB was now considered; however, there was no history of

constitutional/reactivation of TB symptoms (fever, night sweats, weight loss, and hemoptysis) or family history of TB, which we now specifically enquired regarding. The patient also received a complete course of the BCG vaccination during infancy as part of the national immunization program. No biochemical or clinical evidence of reactivated, milliary, or CNS TB was found except for the raised ESR, which was now in conformity with the histopathology. An interferon-gamma release assay (positive) and human immunodeficiency virus test (negative) were obtained.

The patient was eventually discharged on the 14th post admission day, with the family counseled diligently on the importance of compliance with the prescribed 18-month anti-TB drugs. The patient was commenced on a steroid tapering regimen with isoniazid, rifampicin, pyrazinamide, and ethambutol for the first 2 months and is now on isoniazid and rifampicin. At her 8th-month follow-up, her dystonic hand has resolved to complete normality, and she has regained a complete range of motion in her right limbs [Figures 1b and c]. Follow-up MRI [Figure 3] shows a significant reduction in her tuberculoma size. Her gait has significantly improved to allow for independent activities of daily living. A subsequent follow-up is planned in 6 months, at which point the cessation of anti-TB medication will be considered.

DISCUSSION

TB is a significant global public health concern, especially in underdeveloped nations.[11] According to the World Health Organization (WHO), there were approximately 530,000 TB cases in children, with 74,000 deaths in 2012.[11] The WHO estimates that *Mycobacterium tuberculosis* infects around one-third of the world's population.[14] Despite a highly effective pharmacological treatment regimen – "RIPE" (Rifampicin, Isoniazid, Ethambutol, Pyrazinamide) and the BCG vaccination, which is reported to give continuous protection (in 70–80%) against severe types of childhood TB, TB is a leading cause of death in endemic regions.[7,8] After transmission through aerosol, the infection assumes an asymptomatic/latent course of illness.[5] However, it can reactivate years later as secondary pulmonary TB from immunosuppression, a more common occurrence.[5] Less commonly, the primary infection progresses to progressive primary TB, mainly in the immune-compromised, leading to a progressive lung disease and a highly fatal bacteremia

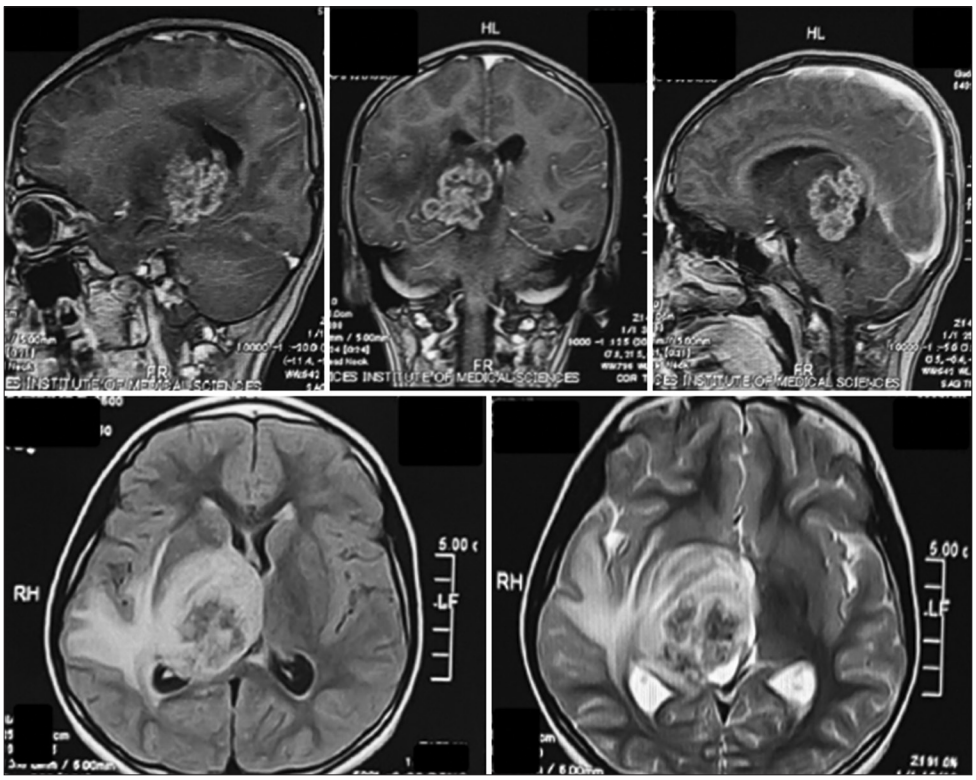

Figure 2: Top row T1 contrast; (bottom left) Fluid attenuated inversion recovery (FLAIR) sequence; (bottom right) T2 sequence. From radiology report – An ill-defined intra-axial heterogeneous lobulated lesion with crenated margins demonstrating patchy contrast enhancement, returning T2 and FLAIR signals involving the right thalamus, posterior limb of the internal capsule, the right half of the midbrain, ipsilateral temporal lobe, hippocampal region, and abutting the splenium of the corpus callosum. There is marked perilesional vasogenic edema. The lesion exerts a mass effect on the body and trigone of the ipsilateral and third ventricles.

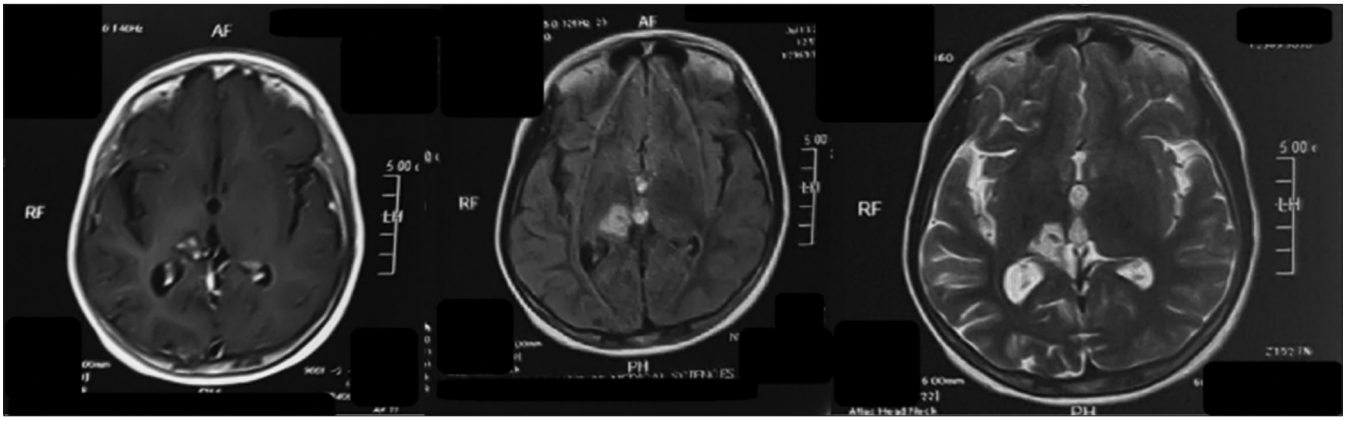

Figure 3: (Left) T1 contrast; (Middle) Fluid attenuated inversion recovery (FLAIR) sequence; (Right) T2 sequence. 8-month follow-up magnetic resonance imaging showing a near complete resolution of the lesion.

dissemination to multiple organs termed miliary TB.[5] The risk of reactivation is approximately 10% (lifetime risk) in the immunocompetent but can be as high as 10%/year in the immunocompromised.[5]

CNS tuberculomas are a rare condition that accounts for approximately 1% of all TB.[9] Unlike CNS tumors, these lesions are slow-growing without immediate destruction to surrounding neural structures, and thus, patients can recover well. Equally, the treatment is paramount as they may rupture in the subarachnoid space, leading to meningitis.[4] CNS tuberculomas are most likely to occur in the immunocompromised. They would be so as part of miliary TB when the disease is disseminated in multiple organs or would exhibit symptoms of reactivation of the primary/latent infection in the form of fever, night sweats, weight loss, or hemoptysis.[5] In the absence of such a sequela, a pediatric immunocompetent vaccinated patient, the "thalamic hand (indicating dystonia) presenting" with ataxic hemiparesis is indeed a rarity, and acute development of such a presentation would more likely point toward an event of ischemic/infarct.[1,3,10] However, as in our case, the gradual development of these symptoms is more in line with a space-occupying lesion such as pediatric tumors, which are far more common. Sahu *et al*.[10] have the first to describe ataxic hemiparesis of a lacunar syndrome secondary to a thalamic tuberculoma in a 65-year-old immunocompetent patient whose chief complaint was similar to our case.[10] The presenting gait in such cases is most likely the result of a distortion to the dentatorubrothalamic tract carrying fibers from the contralateral cerebellum to the ventrolateral nucleus.[10] The associated hemiparesis may likely be from perilesional edema compressing the internal capsule, and the lack of sensory symptoms could result from the spared ventral posteromedial/lateral nuclei.[10] We could find only three cases where an ataxic hemiparesis presentation was due to an intracranial tuberculoma in immunocompetent patients, with our case being the first in a child.[1,10,12] Interestingly, surgical excision for a tuberculoma may be appropriate where there is a life-threatening rise in intracranial pressure from mass effect. Das *et al*.[2] reported such a case of an infratentorial tubercular abscess within the cerebellum which the authors successfully excised in a 5-year-old boy.[2]

According to the British Infection Society Guidelines, children with CNS TB should ideally be managed by a doctor conversant with and knowledgeable about pediatric TB or by a pediatric infectious diseases unit.[13] However, the harsh reality in a third-world resource-constrained country like ours is that such patients are managed by neurosurgeons instead of a multi-disciplinary team like our case due to a lack of such infrastructure for many patients. However, fortunately, the outcome, in this case, may be deemed favorable. Essential learning points are that non-vascular causes should be considered in lacunar syndromes/ataxic hemiparesis dystonia presentations. CNS tuberculoma can be a *de novo* presentation of TB, even without a prior history/clinical symptom with non-specific symptoms/radiology, thus making it a diagnostic challenge. In everyday practice from a neurosurgical principle in such a clinical case – where there is diagnostic uncertainty and a meaningful resection unlikely, a stereotactic biopsy can provide immense clarity and guide management.

CONCLUSION

Without a clinical history or specific symptom, a CNS tuberculoma's clinical and radiological presentation can be non-specific and may indicate a wide range of differentials. A cautious approach in such cases is advised to affirm a diagnosis before definitive treatment.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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

The author(s) confirms 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.

REFERENCES

1. Alarcón F, Tolosa E, Muñoz E. Focal limb dystonia in a patient with a cerebellar mass. Arch Neurol 2001;58:1125-7.
2. Das JM, Sugatha Rao SC, Bhanu Prabhakar R. The successful surgical management of a rare case of infratentorial tubercular abscess in an immunocompetent pediatric patient. Indian J Tuberc 2021;68:529-33.
3. Datta AK, Chakraborty U, Chandra A. The thalamic hand: An enigmatic sequela of thalamic stroke. QJM 2022; 115:239-40.
4. Davis LE. In: Scheld WM, Whitley RJ, Durack DT, editors. Infections of the central nervous system. New York: Elsevier, Raven Press; 1991, 1993. p. 937.
5. Ehrt S, Schnappinger D, Rhee KY. Metabolic principles of persistence and pathogenicity in *Mycobacterium tuberculosis*. Nat Rev Microbiol 2018;16:496-507.
6. Fisher CM. Capsular infarcts: The underlying vascular lesions. Arch Neurol 1979;36:65-73.
7. Katelaris AL, Jackson C, Southern J, Gupta RK, Drobniewski F, Lalvani A, *et al.* Effectiveness of BCG vaccination against *Mycobacterium tuberculosis* infection in adults: A cross-sectional analysis of a UK-based cohort. J Infect Dis 2020;221:146-55.
8. Mashabela GT, de Wet TJ, Warner DF. *Mycobacterium tuberculosis* metabolism. Microbiol Spectr 2019;7:a021121.
9. Mohamud JA, Gu J. Aggressive intracerebral tuberculoma in a 1-year-old child: A case report with literature review. Radiol Case Rep 2020;15:2282-4.
10. Sahu R, Patil TB, Kori P, Shukla R. Isolated thalamic tuberculoma presenting as ataxic hemiparesis. BMJ Case Rep 2013;2013:bcr2013009100.
11. Seddon JA, Shingadia D. Epidemiology and disease burden of tuberculosis in children: A global perspective. Infect Drug Resist 2014;7:153-65.
12. Soto-Cabrera E, Villamil-Osorio LV, Garcia-Luna RC, Carrera-Pineda R. Optochiasmatic tuberculomas as a paradoxical reaction to treatment for meningeal tuberculosis. Rev Neurol 2018;66:286-8.
13. Thwaites G, Fisher M, Hemingway C, Scott G, Solomon T, Innes J, *et al.* British Infection Society guidelines for the diagnosis and treatment of tuberculosis of the central nervous system in adults and children. J Infect 2009;59:167-87.
14. World Health Organization. Staff WHO. Global tuberculosis report 2013. Geneva: World Health Organization; 2013.

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