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Abul Hasan Shadali Abdul Khader
Department of Internal Medicine, Government Kilpauk Medical College, Chennai, India, shadali98@gmail.com

Barath Prashanth Sivasubramanian
Department of Infectious Diseases, University of Texas Health Science Centre, San Antonio, Texas, United States

Diviya Bharathi Ravikumar
Department of Internal Medicine, ESIC, Chennai, India

Nithyalakshmi Rajkumar
, Department of Internal Medicine, Government Kilpauk Medical College, Chennai, India

Gokul Rajakannu
Department of Internal Medicine, Government Kilpauk Medical College, Chennai, India

See next page for additional authors

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Authors

Abul Hasan Shadali Abdul Khader, Barath Prashanth Sivasubramanian, Diviya Bharathi Ravikumar, Nithyalakshmi Rajkumar, Gokul Rajakannu, and Raghavendra Tirupathi

CASE REPORT

Unmasking the Common Thief: A Rare Cause of Isolated Hypoglossal Nerve Palsy

Abul Hasan Shadali Abdul Khader [a,*], Barath Prashanth Sivasubramanian [b], Diviya Bharathi Ravikumar [c], Nithyalakshmi Rajkumar [a], Gokul Rajakannu [a], Raghavendra Tirupathi [d]

[a] Department of Internal Medicine, Government Kilpauk Medical College, Chennai, India
[b] Department of Infectious Diseases, University of Texas Health Science Centre, San Antonio, TX, United States
[c] Department of Internal Medicine, ESIC, Chennai, India
[d] Cure Drug Repurposing Collaboratory, United States

Abstract

Multiple cranial nerve palsies frequently accompany hypoglossal nerve palsy, potentially indicating malignancy, such as lymphoma, nasopharyngeal carcinoma, or metastases. However, when solely the hypoglossal nerve is affected, the causes may involve Chiari malformation, arachnoid cyst, or infectious mononucleosis, suggesting a positive prognosis. Craniocervical junction tuberculosis (TB), is an uncommon cause of isolated hypoglossal nerve palsy and has been reported infrequently in the literature. Craniocervical junction tuberculosis accounts for only 0.5% of TB cases overall and 6% of extra-pulmonary TB cases. We present here one such case of a 17-year-old male of Indian origin with a subacute history of tongue deviation and neck pain. Additionally, the patient reported loss of weight and appetite. The patient had significant posterior cervical lymphadenopathy. Neurological examination revealed findings suggestive of right peripheral hypoglossal nerve involvement. Blood investigations showed lymphocytosis along with an elevated erythrocyte sedimentation rate of 45 mm/h and elevated lactate dehydrogenase levels of 325 U/L. Tuberculin skin testing was positive and sputum acid-fast staining confirmed acid-fast bacilli. Magnetic Resonance Imaging of the cervical spine revealed a soft tissue component in the prevertebral space measuring 3.5× 4.8 cm with a right paraspinal component adjoining the hypoglossal canal with peripheral contrast enhancement. Histological findings on the lymph node showed granulomatous lymphadenitis, suggestive of tuberculosis. The patient was started on 4-drug anti-tubercular therapy consisting of Isoniazid, Rifampicin, Pyrazinamide, and Ethambutol for a period of 18 months. He was subsequently followed up for 6 months till the resolution of palsy. This case emphasizes the importance of thorough evaluation and a meticulous workup to identify the underlying cause of hypoglossal nerve palsy and the importance of considering tuberculosis as a potential cause of isolated hypoglossal nerve palsy in everyday practice.

Keywords: Hypoglossal nerve, Tuberculosis, Malignancy, Cranial nerves, Diagnosis

1. Introduction

The occurrence of hypoglossal nerve palsy is commonly associated with multiple cranial nerve palsies, owing to its proximity to cranial nerves and blood vessels as well as its intricate pathway.[1,2] Despite its rarity, hypoglossal nerve involvement can occur in isolation, with causative factors ranging from neurological anomalies and tumors to post-EBV infection.[1,3] On the far end of the spectrum, tuberculosis in the craniocervical junction is a relatively uncommon presentation, occurring in only 0.5% of all TB cases and 6% of extra-pulmonary TB.[4–6] There exist only a few case reports that describe tuberculosis as the cause of isolated HNP.[1,4,6–8] Untreated cases of tuberculosis are particularly concerning due to complications such as atlantoaxial dislocation [5]. This report presents a case of a 17-year-old patient who presented with deviated tongue and neck pain,

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* Corresponding author.
E-mail address: shadali98@gmail.com (A.H.S. Abdul Khader), barathprashanth18196@gmail.com (B.P. Sivasubramanian), diviyabharathi2612@gmail.com (D.B. Ravikumar), nithyalakshmi.rajkumar97@gmail.com (N. Rajkumar), pavigok32@gmail.com (G. Rajakannu), tirupa@keystonehealth.org (R. Tirupathi).

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further contributing to the corpus of research on this topic.

2. Case report

A 17-year-old adolescent male with an Indian origin presented with a gradual onset of neck pain over the course of one month accompanied with neck stiffness. The pain aggravated with neck movements. Additionally, the patient reported loss of weight and appetite. There was no history of febrile illness, trauma, or recent travel. The patient had significant posterior cervical lymphadenopathy on general examination. A neurological examination indicated a right-ward deviation of the tongue with associated muscle wasting and fasciculations, which is suggestive of significant right peripheral hypoglossal nerve involvement.

The results of the blood investigations (Table 1) revealed anemia with lymphocytosis. However, routine biochemistry and glucose levels were normal. The erythrocyte sedimentation rate was found to be elevated at 45 mm/h, and lactate dehydrogenase levels were also high at 325 U/L. The patient was evaluated for immune compromising conditions, specifically HIV. HIV 1/2 antigen and antibody testing yielded negative results. Given the endemicity of tuberculosis in India and the accompanying constitutional symptoms of weight and appetite loss, evaluation of tuberculosis was considered. As part of the workup, acid-fast bacilli were identified in the sputum, indicating active pulmonary TB. Also, tuberculin skin testing was significant with 11 mm of induration, indicating exposure to tuberculosis. Nevertheless, the chest X-ray and cervical spine X-ray were normal. The magnetic resonance imaging of the cervical spine with gadolinium (Fig. 1) revealed a soft tissue component in the prevertebral space anterior to C2 with a maximum thickness of 1.8 cm and a right paraspinal component measuring 3.5 × 4.8 cm, abutting the right hypoglossal canal with peripheral contrast enhancement. A computed tomography-guided fine needle aspiration of the lesion was planned but was deferred due to the lack of patient consent. Fine-needle aspiration of the lymph node showed granulomatous lymphadenitis with caseation, pointing at tuberculosis.

Table 1. Blood investigations at the time of presentation.

Test	Value
Hemoglobin	9.3 g/dL
White Blood count	9.9×10^9/L
Platelet count	330×10^9/L
HIV/VDRL/HbsAg/HCV/EBV	Negative
ESR	45 mm/h
LDH	325 U/L
Blood Urea Nitrogen	20 mg/dL
Creatinine	1.0 mg/dL
AST	35 U/L
ALT	32 U/L
ALP	55 U/L
Bilirubin	1.1 mg/dL
Albumin	3.9 g/dL
Sodium	137 mEq/L
Potassium	4.0 mEq/L
Calcium	9.6 mg/dL
Anti-HCV	Negative
HbsAg	Negative
HIV 1/2 Serology	Negative
ANA	Negative
ANCA IFA Pattern	None detected
ANCA IFA Titer	1:10
Anti MPO Ab	5 AU/mL
Anti PR3 Ab	6 AU/mL
Serum ACE	14 U/L
CRP	2 mg/L

Serum ACE- Serum angiotensin-converting enzyme.

We ruled out other possible differentials, such as infections like EBV and encephalomyelitis, structural abnormalities like chiari malformation and arachnoid cyst, tumors, and autoimmune diseases such as vasculitis and sarcoidosis (negative ANA, ANCA, Anti PR3, Anti MPO, and low serum ACE levels). Based on clinico–radiological correlation, the final diagnosis was made as craniocervical tuberculosis with right lower motor nucleus hypoglossal nerve palsy. The patient was administered anti-tubercular therapy consisting of isoniazid, rifampicin, pyrazinamide, and ethambutol along with pyridoxine supplements for 18 months. We advised the patient to follow up in the outpatient department every 6 months. On a follow-up visit after six months, the tongue deviation resolved as shown in the figure (Fig. 2).

3. Discussion

Hypoglossal nerve palsy (HNP) commonly occurs with other cranial nerve palsies, mainly due to the nerve's close proximity to several other critical anatomical structures along its course.[2] Studies have emphasized the etiological importance and favourable prognosis in isolated HNP.[6,9] Cranio-cervical junction tuberculosis is one such condition that spreads retrograde, reaching the cranio-vertebral joints and destroying bony and cartilaginous structures leading to cervico-medullary neural compression and occipital-cervical or atlantoaxial instability.[5] The most likely mechanism responsible for nerve palsy is the involvement of the hypoglossal nerve within the skull by tuberculous granulation tissue.[1] This is a relatively uncommon presentation of tuberculosis with only a few case reports in the world literature.[1,4,8,10,11]

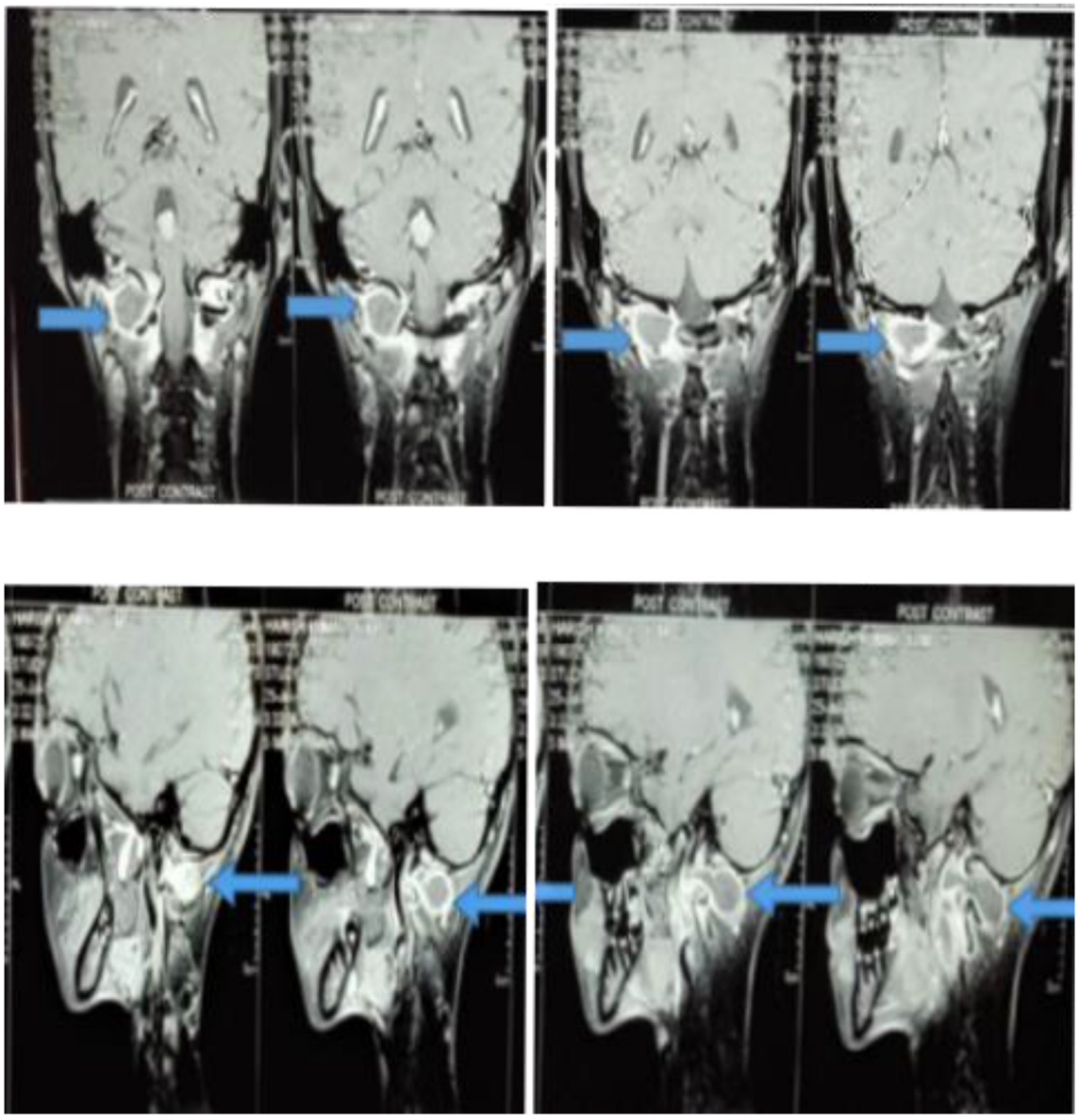

Fig. 1. MRI of the cervical spine showing soft tissue component in the prevertebral space in front of the 2nd cervical vertebrae with a maximum thickness of 1.8 cm with peripheral contrast enhancement.

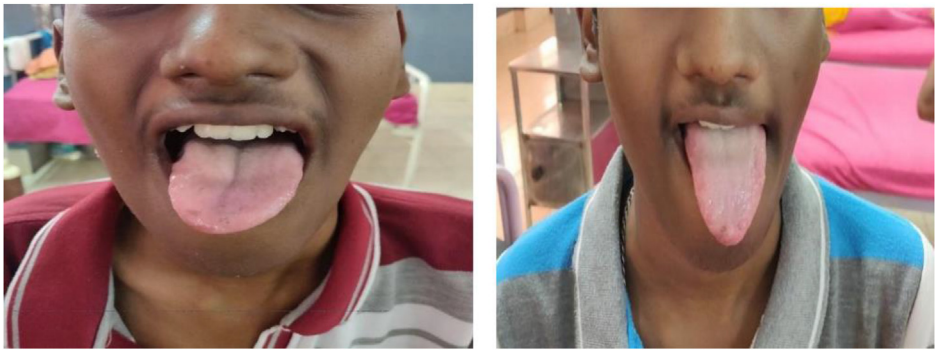

Fig. 2. First image showing deviation of the tongue to the right side before treatment; Second image showing resolution of the deviation after the treatment.

The most common symptoms are occipital and neck pain along with stiffness. They can be non-specific and indistinct in early stages, resulting in a diagnostic delay.[6] In advanced cases, symptoms such as spastic quadriparesis, dysphagia, sensory and sphincter disturbances may be observed.[1] Our literature review revealed neck pain and stiffness of varying intensity in all cases except for the instance described by Jena et al., in 2018, where the patient had a change in voice and tongue deviation.[1,4,8,10,11] In some severe cases, quadriparesis and paraparesis were also observed.[8,10] In our report, the patient experienced neck pain and stiffness for one month, along with loss of weight and appetite. This is in contrast to existing literature[1,4,8,10,11] and emphasizes the importance of focusing on clinical history for such a challenging diagnosis.

The diagnosis of cranio-cervical tuberculosis is typically confirmed via histological examination, demonstrating the presence of granulomatous tissue with or without caseation necrosis.[5] In all reviewed cases,[1,4,8,10,11] the erythrocyte sedimentation rate was elevated but had no correlation with symptom severity or prognosis. Thus, ESR could also be a valuable diagnostic tool in cases presenting with this condition. Tissue culture and pathological examination, along with AFB staining, have been employed to confirm the diagnosis of TB in most cases. However, in some cases, such as our own, initiation of treatment is based on clinical–radiologic correlation.[8] Imaging features commonly showed a typical soft tissue component in the suboccipital region/base of the skull with contiguous extension to the hypoglossal canal, or atypical complex involvement of C1 and C2 vertebrae with involvement of adjoining areas in the base of the skull. The lesions typically had uniform peripheral enhancement in all cases, except for the case described by Chakraborty et al.[1,10,11] In severe cases, destruction of the atlas and axis vertebrae with prevertebral abscess causing spinal cord compression have also been reported.[8]

The management options for tuberculosis affecting the craniocervical junction are subject to variation in practices. While some experts advocate for conservative management in all cases, others suggest surgical intervention based on the severity of the condition.[6,12] Studies have indicated that individualized treatment strategies, based on clinical presentation and radiological findings, are necessary for successful outcomes. Furthermore, a prolonged anti-tubercular treatment (ATT) for 18 months is essential.[6,12,13] It is noteworthy that patients with fixed atlantoaxial dislocation who can perform their daily activities may not require surgery. When surgical approach is required, anterior decompression with posterior stabilization can be performed either through an anterior transcervical or transoral approach.[6]

4. Conclusion

The diagnosis of isolated hypoglossal nerve palsy caused by cranio-cervical tuberculosis is very complex necessitating a high degree of diagnostic ability. TB of the craniocervical junction can manifest without any systemic symptoms, and if not detected promptly and treated appropriately, can result in the destruction of the most mobile segments of the spine. This case report further emphasizes the significance of identifying patients from endemic regions who are at a higher risk, and the rationale for TB workup in such patients. In conclusion, it is crucial to note that despite its self-limited nature and favorable clinical prognosis, isolated hypoglossal nerve palsy should always be approached with the utmost care and attention to detail.

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Consent

Informed consent was obtained from the patient and the parents.

Conflicts of interest

None.

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