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Case Report

Tuberculous rhombencephalitis presenting with pseudo-spinal pattern of sensory loss

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ABSTRACT

Tuberculous rhombencephalitis is an uncommon presentation of intracranial tuberculosis. A 57 year old female, presented with central positional vertigo and features of left 5^{th} and 7^{th} cranial nerve palsy, with a sensory dermatomal level to pain and temperature and diminished vibration sense on left side. On evaluation, was found to have multiple discrete and conglomerate ring enhancing lesions in pons with normal MRI of spine. After extensive evaluation with Magnetic Resonance Spectroscopy, Cerebrospinal fluid and blood investigations and Mantoux test, possible tuberculosis rhombencephalitis was considered and was started on antitubercular treatment, to which she responded well. Our patient had an atypical presentation with sensory level in trunk, which has not been reported in literature. Hence we describe this case and review the neuroanatomical substrates involved. A high index of suspicion is always needed, while dealing with atypical presentations.

1. Introduction

Rhombencephalitis refers to inflammatory diseases affecting the rhombencephalon or hind brain, which consists of brainstem and cerebellum. It has diverse etiologies, which can be broadly classified as infectious, autoimmune and paraneoplastic causes.[1–3] Tuberculous rhombencephalitis is rare and accounts for 2% of cases of rhombencephalitis.[4,5] Even in developing countries endemic for tuberculosis, it is an uncommon presentation and comprises only 2.5- 8% of intracranial tuberculomas.[6–9] Here we describe a case of tuberculous rhombencephalitis in a 57-year-old female, who presented with a sensory dermatomal level in trunk resembling spinal cord pathology. It is essential to know about atypical presentations, since early diagnosis and treatment is key to better outcome in rhombencephalitis.

2. Case Report

A 57 year old right handed female, presented with intermittent brief episodes of positional objective vertigo of 4 months duration, without any associated tinnitus, hearing loss, nausea or vomiting. 2 months later, she noticed reduced sensation over left side of her nose, such that she was not able to feel the temperature of water, while washing face. She also complained of numbness of her left lower limb, which started insidiously over left sole and gradually ascended up till her upper abdomen over a period of 2 months. Around the same time, her daughter noticed mild deviation of angle of mouth to right side and reduced clarity of speech. She did not complain of any difficulty while closing the eye.

She denied any history of headache, blurring of vision, diplopia, vomiting, dysphagia, incoordination or weakness of limbs, bowel or bladder complaints, autonomic symptoms or fluctuation of symptoms. Also she did not have

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any fever, cough, weight loss, joint pain, malar rash, oral or genital ulcers or uveitis. Her past history was notable for hypothyroidism, for which she was on regular treatment.

On examination, she was alert and well oriented, afebrile, and her vital parameters were stable. She had reduced pain and temperature sensation over left side of her nose, suggestive of left 5^{th} nerve palsy and deviation of angle of mouth to the right side, with incomplete closure of left eye and reduced wrinkling over left forehead, suggestive of left 7^{th} nerve lower motor neuron palsy. Other cranial nerves were intact. She also had mild dysarthria and left sided dysmetria. On sensory system examination, she had marked reduction (~50%) of pain and temperature sensation on left lower limb and left side of trunk upto T6 level and mild reduction (~20%) of pain and temperature over patchy areas in right lower limb till L1 level. Vibration sense was reduced till anterior superior iliac spine on left side and medial malleolus on right side. Rest of the central nervous system examination and systemic examination were normal. There was no papilledema, motor weakness or neck rigidity and head impulse test was negative.

Based on the clinical features, possibility of cooccurrence of a pontine and spinal cord lesion was considered. Magnetic resonance imaging (MRI) brain revealed multiple discrete and conglomerate ring enhancing lesions in pons, with perilesional edema in brain stem and bilateral middle cerebellar peduncles. The lesions were isointense on T1 weighted sequence and iso to hypointense on T2 weighted sequence. MRI of the spine was normal.(Figures 1 and 2A,B) Nerve conduction studies of upper and lower limbs were also normal.

Based on the imaging findings, possibility of infectious, neoplastic, metastatic, and autoimmune etiologies were considered and was evaluated extensively. Routine blood investigations showed ESR of 52 mm/hr and normal liver and renal function tests and metabolic parameters. She was negative for HIV, VDRL, Hepatitis B and C infections. Serum galactomannan and blood cultures were negative.

Cerebrospinal fluid examination(CSF) showed cell count of 34/mm^3, 98% lymphocytes, protein 68 mg/dl, glucose 62 mg/dl (corresponding blood sugar-100mg/dl.) and high ADA of 18 IU/L. Gram stain, India ink stain, Ziehl Neelsen stain, bacterial culture, malignant cytology and PCR for mycobacterium tuberculosis were negative.

Chest radiograph showed few calcification specks and Computed tomography (CT) scan of chest with contrast showed ground glassing with septal thickening and few small mediastinal lymph nodes suggestive of infectious etiology. She could not bring out sputum for testing and bronchoscopy and lavage could not be done. Her PPD test (purified protein derivative, Mantoux test) was strongly positive (20 mm).

Work up for autoimmune causes included ANA, ANA profile, HLA B51, anti TPO, C ANCA, P ANCA, serum

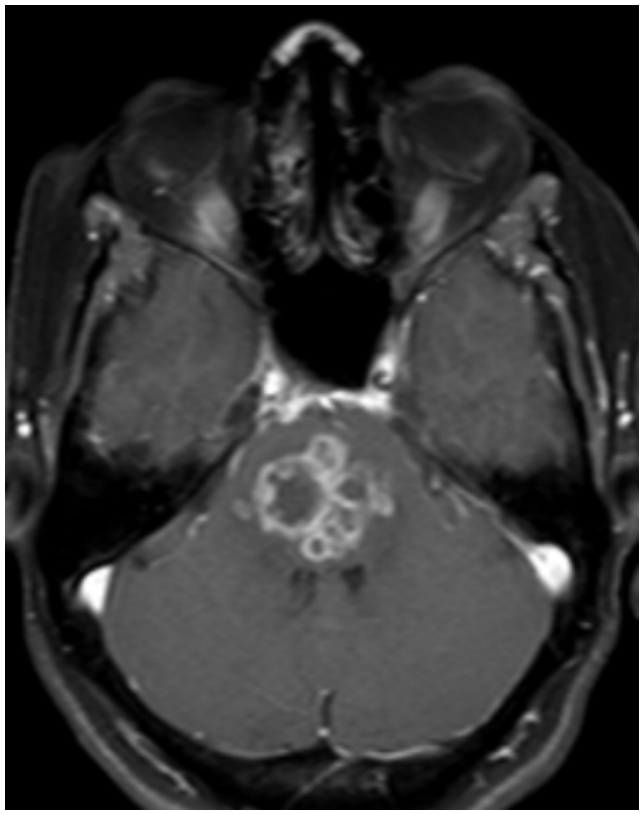

Fig. 1: MRI Brain, Axial post gadolinium contrast T1 Weighted sequence, showing multiple discrete and conglomerate ring enhancing lesions in pons.

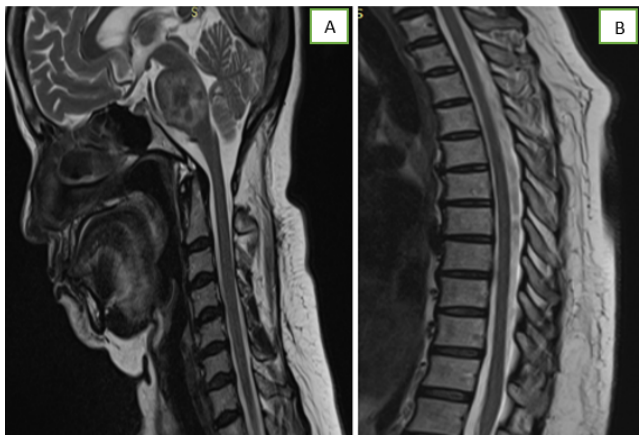

Fig. 2: A,B: MRI cervical spine and dorsal spine, Sagittal T2 weighted sequence, shows normal cervical and dorsal spine.

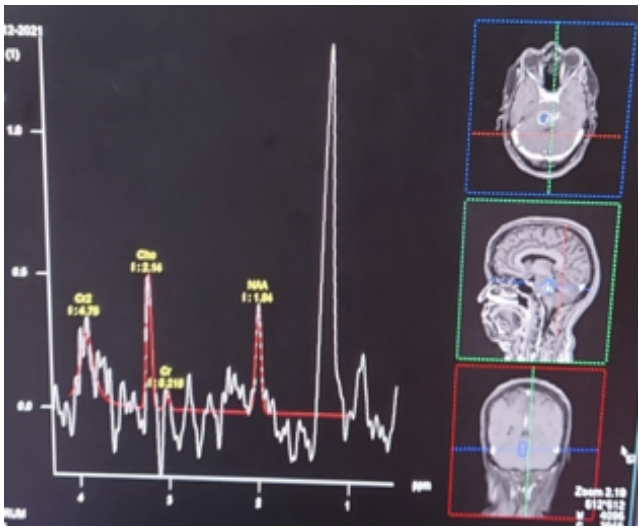

Fig. 3: MRS showing lipid peak at 1.3 ppm.

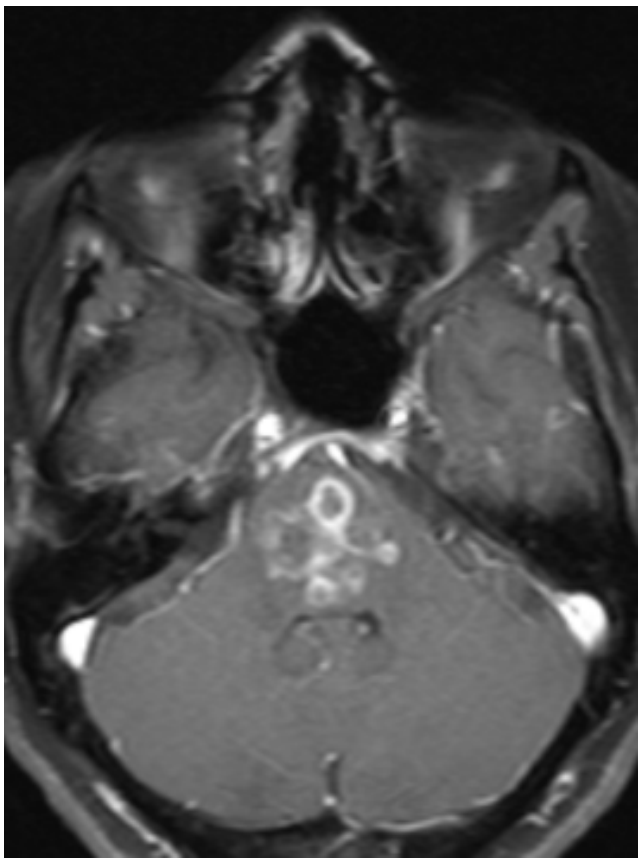

Fig. 4: MRI Brain, Axial T1 Weighted post gadolinium contrast sequence, 6 weeks after initiation of treatment, showing mild reduction in size of lesion.

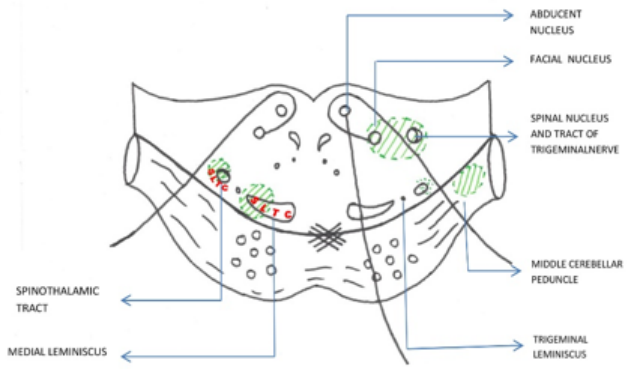

Fig. 5: Pictorial representation of sensory dermatomal lamination of medial leminiscus and spinothalamic tract, in pons.[10,11] Green shaded area indicates lesions in patient. S- Sacral dermatome, L-Lumbar, T-Thoracic, C- Cervical.

angiotensin converting enzyme, IgG4, rheumatoid factor and were negative. She was also extensively evaluated for neoplastic and paraneoplastic causes with tumour markers (Carcinoembryonic antigen, Alphafetoprotein, CA-125, CA 19-9), sonogram of abdomen and pelvis, whole body PET-CT scan and serum paraneoplastic panel and was found to be negative.

Magnetic resonance spectroscopy (MRS) of the lesion showed lipid peak at 1.3 ppm.(Figure 3) In view of imaging findings with MRS, positive PPD test and CSF picture, a possibility of tuberculous rhombencephalitis was considered and she was started on antituberculous treatment with rifampicin, isoniazid, pyrazinamide and ethambutol, with dexamethasone, which was tapered and stopped over 4 weeks.

3 weeks after the initiation of treatment, her facial deviation, left dysmetria and sensory abnormalities of right lower limb resolved. However, vertigo and numbness of left side of face and left lower limb persisted. MRI brain done 6 weeks after the initiation of treatment showed mild reduction of size of lesions, with no new lesion. (Figure 4). 3 months after starting treatment, vertigo and numbness of face and left lower limb also improved significantly.

3. Discussion

Rhombencephalitis is defined as inflammatory diseases affecting rhombencephalon or the hind brain, which is composed of brainstem and cerebellum.[1–5] It was first described by Edwin Bickerstaff and Philip Cloake in 1951.[12] It can be due to multiple etiologies broadly divided into infectious, autoimmune and paraneoplastic causes. The common infections include listeriosis, enterovirus and herpes virus.[1–3] In a case series by Moragas etal, tuberculosis accounted for 2% of all cases.[4] Most common autoimmune causes are Behcets disease, systemic lupus erythematosus and relapsing polychondritis.[1–3] Many of

these conditions are potentially serious and life threatening and leave behind severe sequelae. So early etiological diagnosis and institution of appropriate treatment is crucial. The clinical manifestations depend on etiology and include fever, alteration of consciousness, cerebellar symptoms, cranial nerve palsies and long tract signs.

Tuberculous rhombencephalitis is an uncommon manifestation of intracranial tuberculosis and accounts for 2.5- 8% of all intracranial tuberculomas.[6–9] The radiological features of tuberculous rhombencephalitis are tuberculomas involving parenchyma of hind brain, which can be isointense to hypointense on T1 weighted images and hypointense to isointense or central hyperintensity with a hypointense rim on T2W images, depending on stage of maturation, with edema visualised as hyperintensity on T2 /FLAIR sequences. Involvement of basal cisterns with leptomeningitis also may be seen. On MRS, prominent lipid peak at 1.3 ppm, is characteristic of tuberculoma.[13,14]

Our patient had central positional vertigo, along with brain stem features such as reduced pain and temperature sensation of left side of nose, left lower motor neuron facial palsy and left sided dysmetria. These could be explained by the pontine lesion, involving left spinal nucleus of trigeminal nerve, left facial nerve nucleus or nerve fascicles and left middle cerebellar peduncle.

Our patient had unusual symptoms in form of sensory level in trunk, for which no culprit spinal cord lesion could be found in MRI spine. The sensory dermatomal level in trunk in brain stem lesions has been described in literature. Wang et al reported a case of left pontine infarction with loss of spinothalamic sensations over right side of face and till T4 level in trunk on right side.[10] Leifer et al demonstrated a case of left pontine traumatic lesion with loss of pain and temperature sensation below T4 dermatomal level on contralateral side.[15] These case reports suggest that the sensory dermatomal lamination in spinothalamic tract is preserved upto brain stem level and they speculate that the somatotopic arrangement in spinothalamic tract in brain stem is similar to the arrangement in spinal cord, with sacral fibres placed laterally and cervical fibres arranged medially.[10,15] Hence a lesion involving the lateral part of spinothalamic tract can produce a sensory level in trunk on the contralateral side, as seen in our patient (Figure 5). Apart from a definite sensory level on left side, our patient also had patchy involvement of pain and temperature on right lower limb, which could be due to patchy lesion or edema involving left spinothalamic tract.

Lee et al reported a case of medullary infarct with loss of medial leminiscal sensations only in the contralateral lower limb.[11] The sensory dermatomal lamination of medial leminiscus in medulla is anteroposterior with sacral fibres placed anteriorly and cervical fibres placed posteriorly. In pons, the somatotopic arrangement of medial leminiscus is same as that of spinothalamic tract with sacral fibres placed laterally and cervical fibres placed medially.[11] Hence a lesion affecting the lateral part can cause loss of posterior column sensations on the contralateral lower limb, as seen in our patient.(Figure 5)

The extensive work up did not yield any secondary causes like autoimmune, neoplastic, paraneoplastic causes or evidence of other infections in our case. CSF picture, MRS showing lipid peak, strongly positive PPD test and response to antituberculous treatment point to tubercular etiology, which is highly prevalent in India.

To conclude, rhombencephalitis is an entity with diverse etiologies and the key to better outcome is early diagnosis and prompt treatment. Tuberculous rhombencephalitis is an uncommon manifestation of intracranial tuberculosis and in addition, our patient had a unique presentation with sensory level in trunk, which has not been reported in literature. A high index of suspicion is always needed when dealing with atypical presentations.

4. Conflict of Interest

The authors declare that there are no conflicts of interest in this paper.

5. Source of Funding

None.

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