Case Report

Tumefactive Multiple Sclerosis: A Mimicker of Intracranial Space-Occupying Lesions

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Abstract: A chronic disease of CNS, Multiple sclerosis is characterized by inflammation and demyelination with axonal injury affecting upper motor neurons of brain or spinal cord. Tumefactive multiple sclerosis is a rare form of MS which mimics intracranial tumor-like space-occupying lesions on radiography. A 25-year old gentleman presented 2-month history of gradual onset difficulty in walking, slurring of speech and double vision. The patient had an ataxic gait accentuated by tandem walking with tendency to fall towards right and a negative Romberg's sign. There was dysarthria and signs of cerebellar lesion present on the right side. Muscle tone was normal in upper limbs but increased (spasticity) in lower limbs bilaterally. Power was normal but deep tendon reflexes were exaggerated in all 4 limbs with bilateral up-going plantar reflex. MRI scan of the brain demonstrated tumour-like tumefactive lesions in the right cerebellum, right pons and the left subcortical region with no post-contrast enhancement of these lesions. MRI of whole spine demonstrated subtle hyperintense signals in the cervical spine. He had a past history of transverse myelitis 2 years ago with full clinical recovery with oral steroids. He was diagnosed with Tumefactive Multiple Sclerosis and started on corticosteroid therapy resulting in dramatic improvement.

Keywords: Multiple sclerosis, Tumefactive lesions, Demylination, MRI scan, Oral steroids, Radiography.

INTRODUCTION

A chronic inflammatory disease of the central nervous system, Multiple Sclerosis (MS) is characterized by inflammation and demyelination with axonal injury [1]. Considered to be an autoimmune disorder, the exact etiology of MS is undetermined [2]. It is postulated that MS is mediated by autoreactive Th-1 and Th-17 cells leading to proinflammatory IL-1,IFN- γ and IL-17 cytokine production causing generation of metalloproteinases and destruction of the blood–brain barrier permiting the Th cells to attack the CNS [2]. Usually affecting patients between 20 to 40 years of age, MS is two-fold more common in females as compared to males [3]. MS is the leading cause of disability in young adults in the developed world [4]. Based on the clinical features and course, MS has been categorized into 4 classes: Relapsing remitting MS (episodic nature with acute deterioration of neurologic functioning with partial or complete recovery but no apparent disease progression), Primary Progressive MS (steady disease worsening from the start with no distinct remission or relapse), Secondary Progressive MS (progressive disease with or without relapse) and Progressive Relapsing MS (steady disease worsening from the start with occasional relapses) [3, 5].

Tumefactive Multiple Sclerosis (TMS) is a rare form of MS which mimics intracranial tumor-like space-occupying lesions on radiography usually having a diameter ≥2 cm [6]. Clinical features of TMS are variable depending on site and size of the lesions. The mainstay of diagnosis are Magnetic Resonance Imaging (MRI) scan of brain and spinal cord, Positron Emission Tomography (PET) scan and Cerebo-Spinal Fluid (CSF) analysis.1 Estimates show that TMS affects approximately 3 people per million yearly in the general population and accounting to 1 per 1000 patients of MS [7, 8]. In patients with no history of previous MS, diagnosing TMS is challenging and may be misdiagnosed [7]. The differential diagnosis to consider in a patient with TMS include intracranial space-occupying lesions such as brain tumors and cerebral abscesses [8].

CASE REPORT

We report the case of a 25-year old gentleman who presented with 2-month history of difficulty in walking, slurring of speech and double vision. It was of insidious onset and progressively worsening causing the patient significant impairment in doing routine activities. There was no history of trauma, fever, headaches, loss of consciousness, urinary or fecal incontinence, hearing disturbance, syncope, dysphagia, numbness or tingling. There was a past history of weakness of both lower limbs 2 years ago that was diagnosed as transverse myelitis on MRI spine and reported full clinical recovered after taking oral steroids. However, the patient discontinued the treatment on recovery and did not follow-up. He worked as a cook but had given up 1 month

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ago due to his illness. He was unmarried and had no history of premarital affairs. He did not smoke or use illicit drugs. There was no family history of any similar disorder. He was seen by a local GP who suspected a posterior circulation stroke and referred the patient to Neurology for brain imaging.

On examination, the patient had an ataxic gait accentuated by tandem walking with tendency to fall towards right and a negative Romberg's sign. There was dysarthria but higher mental functions were intact. Signs of cerebellar lesion were present on the right: impaired dysdiachokinesia, dysmetria, intention temor, past pointing and pendular knee jerk. A horizontal nystagmus was noted which worsened on looking towards right. Pupils were round, equal and responsive to light bilaterally with normal extraocular movements. All cranial nerves were intact. Muscle tone was normal in upper limbs but increased (spasticity) in lower limbs bilaterally. Power was normal but deep tendon reflexes were exaggerated in all 4 limbs with bilateral up-going plantar reflex. There was no sensory loss. Examination of cardiac, respiratory and gastrointestinal systems was unremarkable.

On investigation, CBC, ESR, LFTs, RFTs and urinalysis were normal. Serologies for TPHA, HBV, HCV and HIV were negative. Serum B12, HbA1c and TSH were normal. MRI scan of the brain and spinal cord demonstrated tumour-like tumefactive lesions in the right cerebellum (Fig. **1**), right pons (Fig. **2**) and the left subcortical region (Fig. **3**). There was no post-contrast enhancement of these lesions. MRI of whole spine was done which demonstrated subtle hyperintense signals in the cervical spine only (Fig. **4**). The patient did not consent for lumbar puncture, therefore a CSF analysis could not be done. Based on history, examination and radiologic findings he was diagnosed with Tumefactive Multiple Sclerosis and started on intravenous methylprednisolone 1000mg per day for 5 days resulting in dramatic improvement. Subsequently he was prescribed oral azathioprine and oral prednisolone along with calcium and vitamin D supplements, and discharged with a plan to follow-up in OPD.

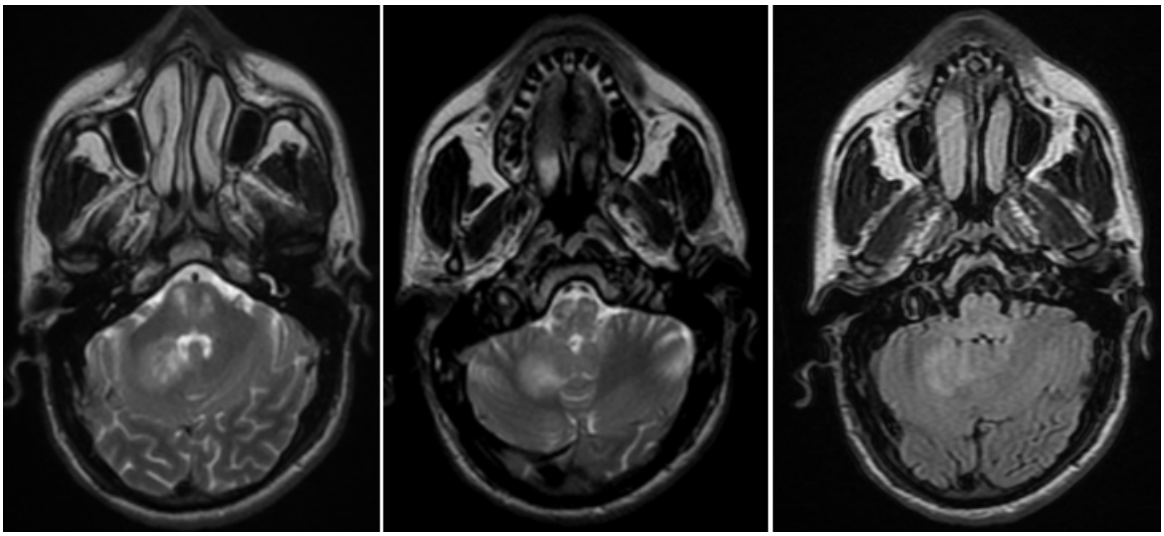

Fig. (1). Abnormal Tumor-Like T2WI/FLAIR Hyperintense Signals Seen in the Right Cerebellum.

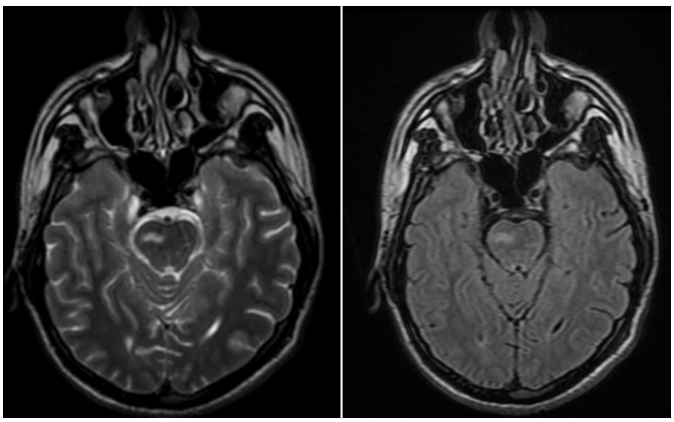

Fig. (2). Abnormal T2WI/FLAIR Hyperintense Signals Seen in the Right Pons.

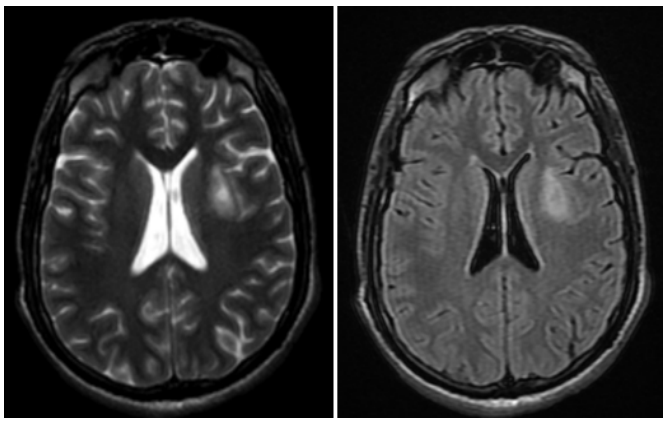

Fig. (3). Abnormal Tumor-Like T2WI/FLAIR Hyperintense Signals seen in the Left Subcortical Region.

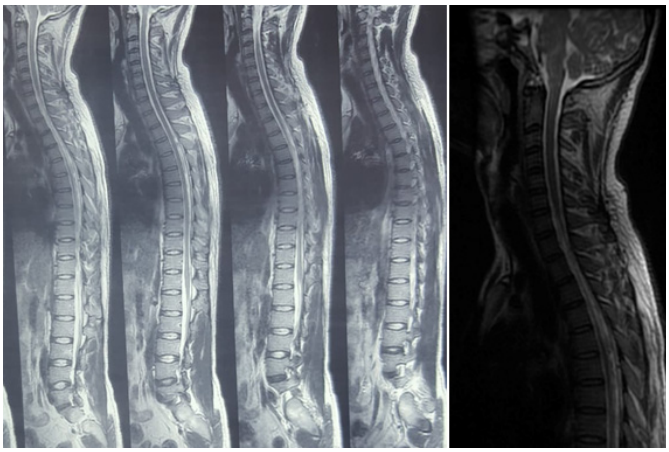

Fig. (4). Abnormal T2WI Hyperintense Signals Seen in the Cervical Spine.

DISCUSSION

In routine clinical practice, MS is diagnosed using the 2017 McDonald criteria which incorporates medical history, neurologic examination and MRI findings in addition to oligoclonal bands in CSF to evaluate both dissemination in time and dissemination in space [9]. The 2017 Mcdonald Criteria has been shown to have high sensitivity (100%) but may have a low specificity (13.8%) [10]. Not only helping in differentiating other CNS disorders, MRI scan also aids in treatment response evaluation and disease progression follow-up [11]. Our patient had dissemination in time evidenced by a past episode of transverse myelitis in addition to the present neurologic findings. Dissemination in space was evidenced by the presence of multiple lesions in right cerebellum, right pons, left subcortical region and the cervical spine in our patient. Our patient fulfilled the 2017 McDonald criteria by having 2 attacks and >2 lesions. The lesions on MRI appear as tumor-like >2cm in size, therefore making the diagnosis of Tumefactive MS in our patient. Our patient did not consent for lumbar puncture and therefore CSF could not be assessed for oligoclonal bands.

The first-line treatment of MS is corticosteroids. In mild cases, oral prednisolone may be given. However, moderate to severe cases require intravenous methylprednisolone therapy [12]. In patients not responding to steroids, plasmapheresis or plasma exchange may be done followed by cyclophpshamide or rituximab if required [13]. For relapsing-remitting disease course, various immunomodulator therapies may be prescribed including beta-interferon, glatrimer acetate, natalizumab, ocrelizumab, alemtuzumab, mitoxantrone and fingolimod [14-17]. In addition, patients require symptomatic therapy tailored according to their presentation along with physiotherapy and occupational rehabilitation. The treatment of TMS is not different from MS. The first-line treatment of TMS compromises corticosteroids and azathioprine [12, 13]. In most cases of TMS, a conservative approach with the use of radiological modalities and clinical follow-up is enough [18]. Our patient was started on intravenous methylprednisolone followed by oral azathiorpine and oral prednisolone and showed dramatic improvement. He was tolerating therapy without any adverse effects and was regular with follow ups.

CONCLUSION

In conclusion, Tumefactive Multiple Sclerosis is a rare form of MS which mimics intracranial tumor-like space-occupying lesions on radiography. It is therefore important to diagnose this condition timely and differentiate it from other diagnoses so that proper management may be initiated to improve outcome.

CONFLICT OF INTEREST

Declared none.

ACKNOWLEDGEMENTS

Declared none.

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Received: May 08, 2023 Revised: January 11, 2024 Accepted: January 12, 2024