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CASE REPORT

Severe neurological manifestations of LETM with brainstem involvement: A dual case report

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Abstract

Background: Transverse myelitis (TM) is a rare neurological disorder characterized by inflammation of the spinal cord, which can cause varying degrees of motor, sensory, and autonomic dysfunction. Longitudinally extensive transverse myelitis (LETM) refers to TM that extends over three or more vertebral segments and may be associated with brainstem involvement, leading to complex clinical symptoms.

Case presentation: This case report presents two cases of LETM with concurrent brainstem involvement, both the patients exhibited progressive neurological deficits, including limb weakness, sensory disturbances and cranial nerve related symptoms.

Imaging findings and management: MRI examination of the spine revealed T2 Hyperintense lesions extending over multiple vertebral segments, consistent with LETM. Brain MRI in both cases showed hyperintensities involving the medulla and pons, indicating brainstem involvement. The patients were managed with high-dose corticosteroids and supportive therapy. One of the patients showed marked clinical improvements, while the other required prolonged rehabilitation with partial neurological recovery.

Conclusion: LETM with brainstem involvement represents a severe neurological condition with complex clinical and radiological features. Early diagnosis through MRI and prompt immunosuppressive therapy are essential for improved outcomes.

KEYWORDS
Longitudinally extensive transverse myelitis (LETM), MRI features in LETM, Brainstem involvement in LETM

INTRODUCTION

Myelopathy is any form of spinal cord pathology whereas myelitis refers to an inflammatory or infectious process.[1] Transverse myelitis (TM) is an infectious or inflammatory disorder that affects the spinal cord and typically disrupts the transmission of nerve signals, leading to symptoms such as paralysis, sensory loss, pain, and autonomic dysfunction. When the inflammation spans more than three segments of the spinal cord, it is classified as longitudinally extensive transverse myelitis (LETM).[2] LETM is often associated with more severe outcomes and may have an underlying etiology, including infections, autoimmune diseases, or demyelinating conditions like multiple sclerosis (MS). In rare cases, brainstem involvement may occur, complicating the presentation as well as management. The radiological features in MRI can suggest or help to differentiate the various etiologies of LETM.[3,4]

CASE PRESENTATION

Case 1

A 32-year-old male presented to the emergency department with complaints of rapidly progressive weakness and numbness in both legs, which had developed over the past 72 hours. He also reported dizziness, dysphagia, and blurred vision. The patient had no significant past medical history but had a recent viral upper respiratory infection. On neurological examination, the patient exhibited paraplegia, dysarthria, and dysphagia. MRI of the cervical and thoracic spine was done which showed presence of longitudinally extensive lesions spanning from C2 to T1 and D3 to D7, characterized by hyperintensity on T2-weighted images and hypo-intensity on T1-weighted images. These lesions appeared to involve both gray and white matter of the spinal cord, with no evidence of significant atrophy or structural abnormalities of the surrounding vertebrae. The MRI of the Brain revealed subtle hyperintensities on T2-weighted images in the brainstem, especially in the pons on right side. There was no gadolinium enhancement in the area. These findings were consistent with brainstem involvement in the context of transverse myelitis. There was no significant lesions were noted in other parts of the brain, which helped exclude diagnoses such as multiple sclerosis or other demyelinating diseases. The inference drawn from these findings was that of LETM with brainstem involvement, secondary to an infectious etiology (Figure1).

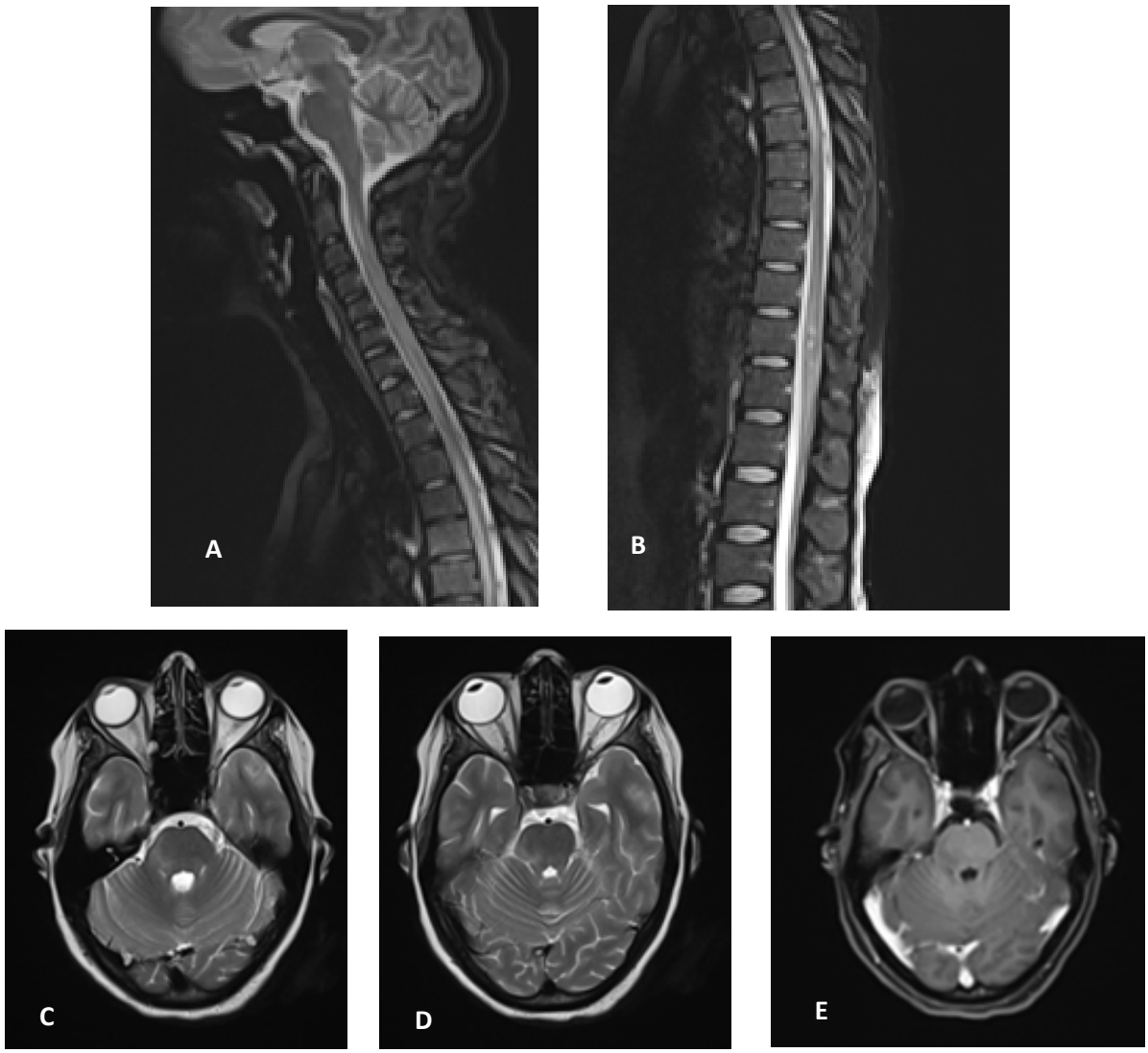

FIGURE 1 **MRI of the cervical and thoracic spine (A and B, STIR sagittal cervical and thoracic, C and D axial T2W, E post contrast axial) showed presence of longitudinally extensive lesions spanning from C2 to T1 and D3 to D7, characterized by hyperintensity on T2-weighted images and hypo-intensity on T1-weighted images without atrophy of the cord. The MRI of the Brain revealed subtle hyperintensities on T2-weighted images in the brainstem, especially in the pons on right side. There was no gadolinium enhancement in the area.**

Case 2

A 42-year-old female presented with acute onset of weakness and sensory loss in both lower limbs, accompanied by bladder dysfunction and loss of deep tendon reflexes. There was no history of trauma, and the patient was previously healthy. The symptoms developed rapidly over the course of 48 hours. Neurological examination revealed signs consistent with myelopathy, including impaired motor strength, decreased sensation below the T10 dermatome, and absent reflexes. The MRI of the spine in this case revealed a longitudinally extensive lesion involving the cervical and thoracic spinal cord, spanning from C3 to T10 with mild expansion of the cord. The lesion appeared hyperintense on T2-weighted images and hypointense on T1-weighted images. The MRI of Brain in this case also showed a lesion in the brainstem, predominantly involving the pons (Left>right), with areas of hyperintensity on T2-weighted images. No significant gadolinium enhancement was observed in the brainstem lesions. The conclusion drawn in this case was also LETM with brainstem involvement – suspected to be of demyelinating etiology (Figure 2).

The patients were managed with high-dose corticosteroids and supportive therapy. the first case patient showed marked clinical improvements, while the second case required prolonged rehabilitation with partial neurological recovery.

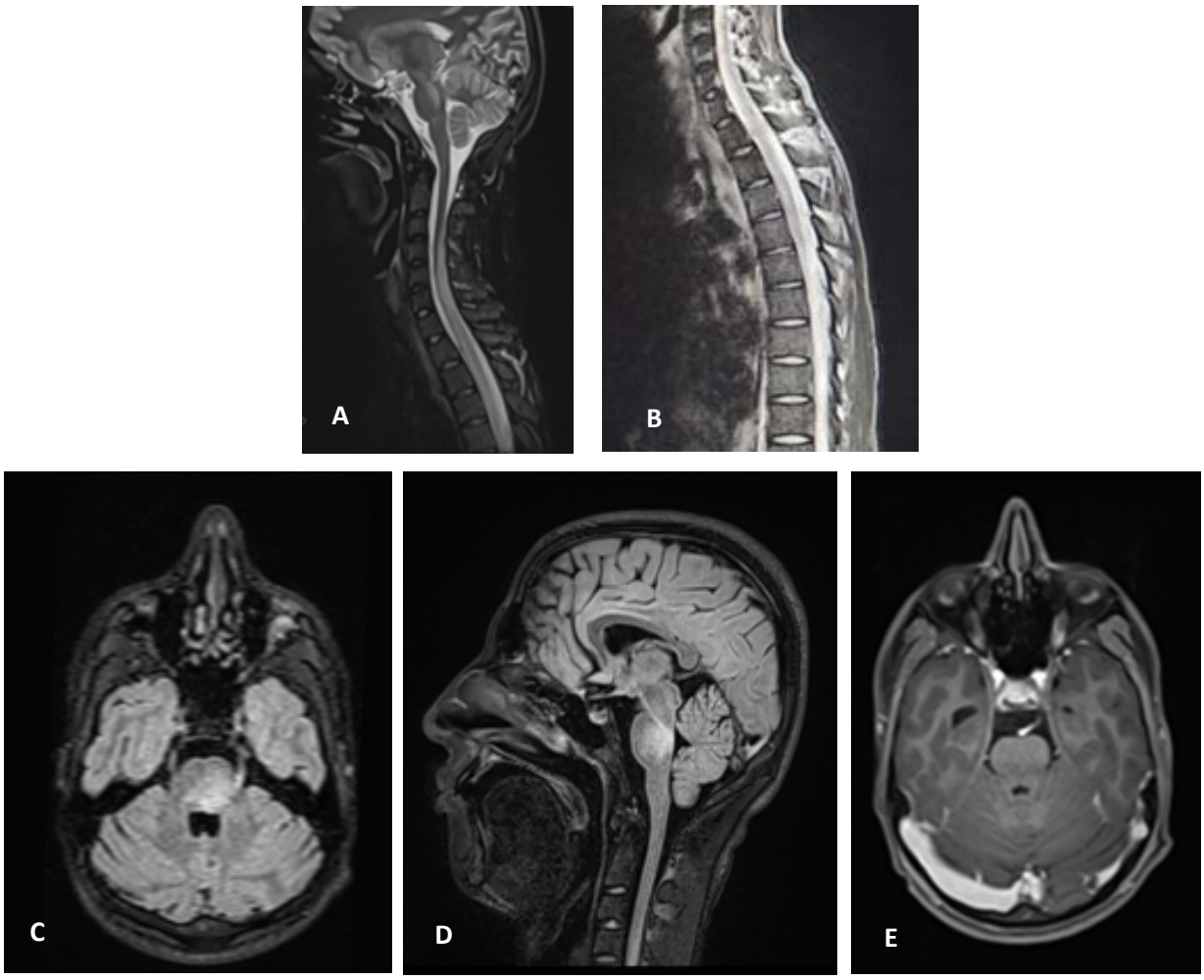

FIGURE 2 The MRI of the spine (A- sagittal STIR cervical , B-Sagittal T2W Thoracic, C and D axial and sagittal FLAIR brain and D-Axial brain post contrast) revealed a longitudinally extensive lesion involving the cervical and thoracic spinal cord, spanning from C3 to T10 with mild expansion of the cord. The lesion appeared hyperintense on T2-weighted images and hypointense on T1-weighted images. The MRI of Brain in this case also showed a lesion in the brainstem, predominantly involving the pons(Left>right), with areas of hyperintensity on T2-weighted images. No significant gadolinium enhancement was observed in the brainstem lesions.

DISCUSSION

Longitudinally extensive transverse myelitis has characteristics of contiguous lesions in the spinal cord and when brainstem involvement presents a diagnostic challenge due to the wide array of differential diagnoses, viral infections, and autoimmune disease neuromyelitis Optica and rarely with multiple sclerosis.[3,4] The clinical presentation is usually with single or multiple attacks of paraparesis or tetra-paresis, sensory deficits and bladder/bowel disturbances and may even progress to respiratory failure.[5] The brainstem involvement, although less common in LETM, may explain the more severe clinical manifestations, including cranial nerve dysfunction, dysphagia, and facial weakness. The detection of such lesions on brain MRI is crucial for accurate diagnosis and management, especially when a concomitant inflammatory or autoimmune disorder is suspected.[7] The MRI findings in our case series reveal characteristic patterns of demyelination and infection in both the spinal cord and brainstem.

Additionally, longitudinal involvement of the spinal cord and brainstem on MRI may help differentiate LETM from other spinal cord pathologies like multiple sclerosis, which typically involves discrete lesions separated by normal-appearing tissue.[8-10]

The following is to be considered in this clinical and radiological settings:

- **Multiple Sclerosis (MS):** Characterized by multiple demyelinating plaques in the central nervous system (CNS). However, MS typically involves fewer than three spinal segments and may show more discrete brain lesions.
- **Neuromyelitis Optica Spectrum Disorder (NMOSD):** This condition is associated with optic neuritis and transverse myelitis but often presents with optic nerve and spinal cord involvement rather than brainstem involvement.
- **Viral Infections:** Such as herpes simplex virus (HSV) or Epstein-Barr virus (EBV) can cause transverse myelitis with or without brainstem involvement, but the rapid progression of symptoms was not typical for viral etiology.
- **Autoimmune Disorders:** Such as systemic lupus erythematosus (SLE) or vasculitis, can cause myelitis but were excluded by the absence of systemic signs of active disease.

  The presence of LETM mandates a prompt search for disease entities like neuromyelitis Optica and myelin oligodendrocyte glycoprotein.

CONCLUSION

LETM with brainstem involvement is a complex neurological disorder requiring high clinical suspicion and advanced imaging techniques for accurate diagnosis. MRI remains the cornerstone of diagnosis, and in cases with brainstem involvement, it is essential for distinguishing between potential etiologies. The radiological findings in this case series underscore the importance of early detection and monitoring of these lesions to guide treatment decisions and improve patient outcomes. Further studies on the long-term prognosis and treatment responses in patients with LETM and brainstem involvement are needed.

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