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A Rare Case of Transverse Myelitis Secondary to Hemophagocytic Lymphohistiocytosis in an Elderly Patient

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Abstract

Background

Transverse myelitis (TM) is a rare neurological condition characterized by spinal cord inflammation leading to motor and sensory deficits. We present a case of TM secondary to hemophagocytic lymphohistiocytosis (HLH) in a patient with diffuse large B-cell lymphoma (DLBCL). Despite initial improvement with corticosteroids and intravenous immunoglobulin, the patient experienced neurological relapse. This case highlights the importance of recognizing TM as a possible manifestation of HLH, the role of CNS imaging, and the potential involvement of DLBCL in spinal cord pathology [1,2,4].

Keywords: Transverse Myelitis; Hemophagocytic Lymphohistiocytosis; Neurology; Transverse Myelitis; Hemophagocytic Lymphohistiocytosis

Background

Transverse myelitis (TM) is a rare neurological condition characterized by inflammation of the spinal cord, often resulting in motor and sensory deficits. This report presents a case of TM secondary to hemophagocytic lymphohistiocytosis (HLH), a severe and life-threatening systemic inflammatory syndrome, in a patient with underlying diffuse large B-cell lymphoma (DLBCL). The case highlights the challenges in diagnosing and managing TM associated with HLH, especially when complicated by potential CNS involvement.

Case Description

A right-handed male in his late 60s presented to the emergency department with sudden onset back pain, lower limb weakness, bladder disturbances, and sensory changes. His initial symptom was isolated back pain experienced while on holiday. Three days later, the patient developed worsening weakness, leading to faecal incontinence. He had a medical history of type 2 diabetes, renal calculi, resected squamous cell carcinoma of the lower limb, cataracts, and a right hip replacement. Functionally independent prior to this incident, he reported no recent fever, infections, or trauma.

The patient was initially discharged home from the emergency department but returned five days later with confusion, drowsiness, and suprapubic tenderness. Although a positive quantiferon test indicated latent TB, this was considered incidental, and no neurological symptoms were linked to TB. Anti-tubercular treatment was initiated by the infectious disease team due to unresolved sepsis without a clear infection source.

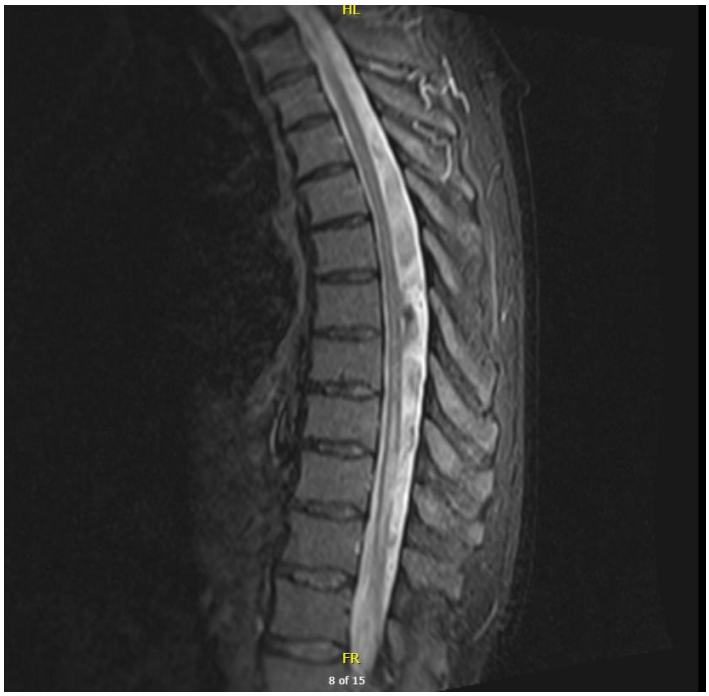

Figure 1: An MRI of the cervical and thoracic spine showed abnormal T2 hyperintensities in the distal thoracic cord from T9 to T11, confirming acute myelitis

Results of Investigations

On neurological examination, cranial nerves and upper limb function were intact, but significant weakness, hyperreflexia, and an extensor plantar response were noted in the lower extremities, with a sensory level at T10. Routine blood tests revealed mild thrombocytopenia. Initially admitted under orthopedics, an urgent MRI of the lumbar spine showed mild to moderate disc changes. Neurology was consulted due to suspicion of TM or acute myelopathy.

The patient underwent a comprehensive workup,

including vasculitic markers and antibodies for MOG and NMO, all returning negative. An MRI of the cervical and thoracic spine showed abnormal T2 hyperintensities in the distal thoracic cord from T9 to T11, confirming acute myelitis. Treatment with intravenous steroids (three days) followed by IVIG (five days) led to significant recovery, enabling the patient to self-mobilize. He was discharged on a tapering dose of oral steroids.

Upon readmission, persistent cytopenias, elevated ferritin levels, and low NK cell activity were identified, meeting the diagnostic criteria for HLH.

HLH Diagnostic Criteria Met

- Fever (temperature > 38.5° C for over seven days)
- Splenomegaly (palpable > 3 cm below costal margin)
- Cytopenia involving two cell lines (hemoglobin < 9 g/dL, ANC < 100/mcL, platelets < 100,000/mcL)
- Hypofibrinogenemia (fibrinogen < 150 mg/dL)
- Hemophagocytosis in bone marrow biopsy
- Elevated serum ferritin > 500 ng/mL (notably 1123.5 ng/mL)

CSF analysis showed elevated protein (0.60 g/L) and WCC (13/cmm), findings consistent with neuroinflammatory involvement in transverse myelitis.

Discussion

The patient's HLH diagnosis was attributed to DLBCL, which can involve the CNS and potentially lead to neuroinflammation. While latent TB was incidentally identified, it was not considered the primary cause of his symptoms. Due to his septic presentation and lack of a clear source of infection, anti-TB treatment was initiated by the infectious disease team. Despite initial improvement, the patient experienced a relapse of neurological symptoms, highlighting the complex and unpredictable nature of HLH [3,6]. CNS hemophagocytosis, as seen in this case, is rare and often associated with severe neuroinflammatory complications [2,5].

DLBCL can involve the CNS, including the spinal cord, leading to neuroinflammation and neurodegeneration. Although primary spinal cord DLBCL is rare, it should be considered in cases of TM with concurrent HLH, as CNS infiltration by lymphoma cells can contribute to neuroinflammatory damage. In this case, while lymphoma cells were not detected in CSF, CNS involvement was suspected based on clinical presentation and imaging [1,3,5].

This case underscores the importance of early recognition and comprehensive management of HLH and associated neurological complications. Given DLBCL's potential to involve the CNS, including the spinal cord, clinicians should maintain a high index of suspicion for TM in similar presentations. Neuroimaging, such as MRI, is crucial in identifying spinal cord involvement, as demonstrated by the high T2 signal observed in this patient's MRI [4,6].

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