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Acute-onset quadriplegia with hyperreflexia

A 79-YEAR-OLD MAN PRESENTED with sudden-onset bilateral weakness in the lower and upper extremities that had started 6 hours earlier. He reported no vision disturbances or urinary incontinence. He was afebrile, with a blood pressure of 148/94 mm Hg, heart rate 98 bpm, and oxygen saturation of 95% on room air.

Physical examination revealed quadriplegia with hyperreflexia, sustained ankle clonus, and bilateral Babinski reflex, as well as spontaneous adductor and extensor spasms of the lower extremities.

Funduscopy was negative for optic neuritis. Results of a complete blood cell count and renal and liver function testing were within normal limits.

Because the patient's presentation raised concern for cervical cord compression, urgent magnetic resonance imaging (MRI) of the cervical spine was performed, with and without contrast. It showed abnormal diffuse T2 hyperintensity beginning at the level of the medulla and extending inferiorly to level C7 (Figure 1). This led to a diagnosis of longitudinally extensive transverse myelitis (LETM).

The patient was admitted to the intensive care unit. Methylprednisolone 1 g was given intravenously once daily for 5 days, with plasma exchange every other day for 5 sessions. A workup for neoplastic, autoimmune, and infectious disease was negative, as was testing for serum aquaporin-4 antibody (ie, neuromyelitis optica immunoglobulin G antibody).

Over the course of 7 days, the patient's motor strength improved, and he was able to walk without assistance. Steroid therapy was tapered, and he was prescribed rituximab to prevent recurrence.

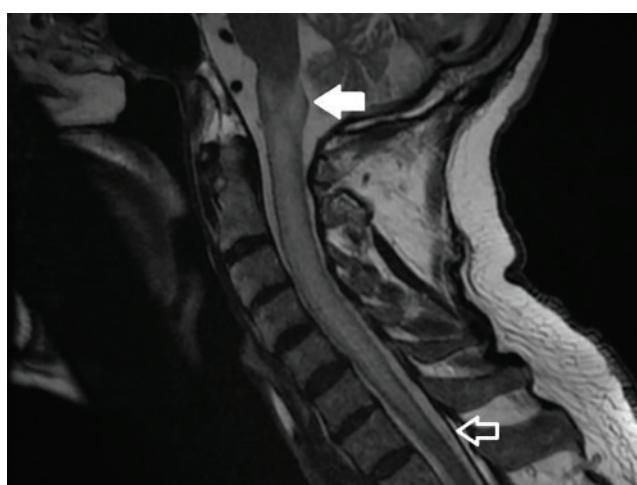


Figure 1. Magnetic resonance imaging of the cervical spine without contrast showed abnormal diffuse T2 hyperintensity beginning at the level of the medulla (solid arrow) and extending inferiorly to the level of C7 (open arrow).

LONGITUDINALLY EXTENSIVE TRANSVERSE MYELITIS

A subtype of transverse myelitis, LETM is defined by partial or complete spinal cord dysfunction due to a lesion extending 3 or more vertebrae as confirmed on MRI. The clinical presentation can include paraparesis, sensory disturbances, and gait, bladder, bowel, or sexual dysfunction.¹ Identifying the cause requires an extensive workup, as the differential diagnosis includes a wide range of conditions²:

- Autoimmune disorders such as Behçet disease, systemic lupus erythematosus, and Sjögren syndrome
- Infectious disorders such as syphilis, tuberculosis, and viral and parasitic infections
- Demyelinating disorders such as multiple sclerosis and neuromyelitis optica

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- Neoplastic conditions such as intramedullary metastasis and lymphoma
- Paraneoplastic syndromes.

In our patient, the evaluation did not identify a specific underlying condition, and testing for serum aquaporin-4 antibody was negative. Therefore, the LETM was ruled an

isolated idiopathic episode.

Idiopathic seronegative LETM has been associated with fewer recurrences than seropositive LETM.³ Management consists of high-dose intravenous steroids and plasma exchange. Rituximab can be used to prevent recurrence.⁴ ■

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