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Teaching NeuroImage: Subacute Quadriparesis From Intramedullary Spinal Cord Infiltrating Glioma With TERT Promoter Mutation

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A 68-year-old man without medical history developed two months of progressive weakness and cervicalgia. Exam showed quadriparesis with T10 sensory level. Spine MRI revealed an expansile intramedullary lesion from obex to T11 with peripheral nodular enhancement (Figure, A–D). Brain MRI, body PET/CT, and broad serum and CSF diagnostics were normal (eTable 1). CSF showed protein 2,505 mg/dl, 0 cells/ul, glucose 88 mg/dl and CSF cell free DNA sequencing identified a pathogenic variant in TERT p.C250T, suspicious for glioma.[1] Thoracic spinal cord biopsy was pursued to exhaust reversible etiologies and revealed infiltrating glioma with TERT promoter mutation (Figure, E–F). Due to progressive quadriplegia, respiratory failure, and poor prognosis, care was directed towards comfort.

Spinal masses are classified as extradural, intradural extramedullary, or intradural intramedullary.[2] Differential diagnosis for intramedullary cord lesions includes demyelination, paraneoplastic myelopathies (e.g. anti-CRMP5), neuro-sarcoidosis, infection, vascular abnormalities (e.g. Dural arteriovenous fistula/malformation), nutritional deficiency, toxic insult, or tumor. While non-invasive diagnostics should be exhausted, definitive diagnosis of neoplastic myelopathy generally requires biopsy. Novel cell free DNA sequencing may complement or eventually supersede certain diagnostics, especially where biopsy is unsafe.

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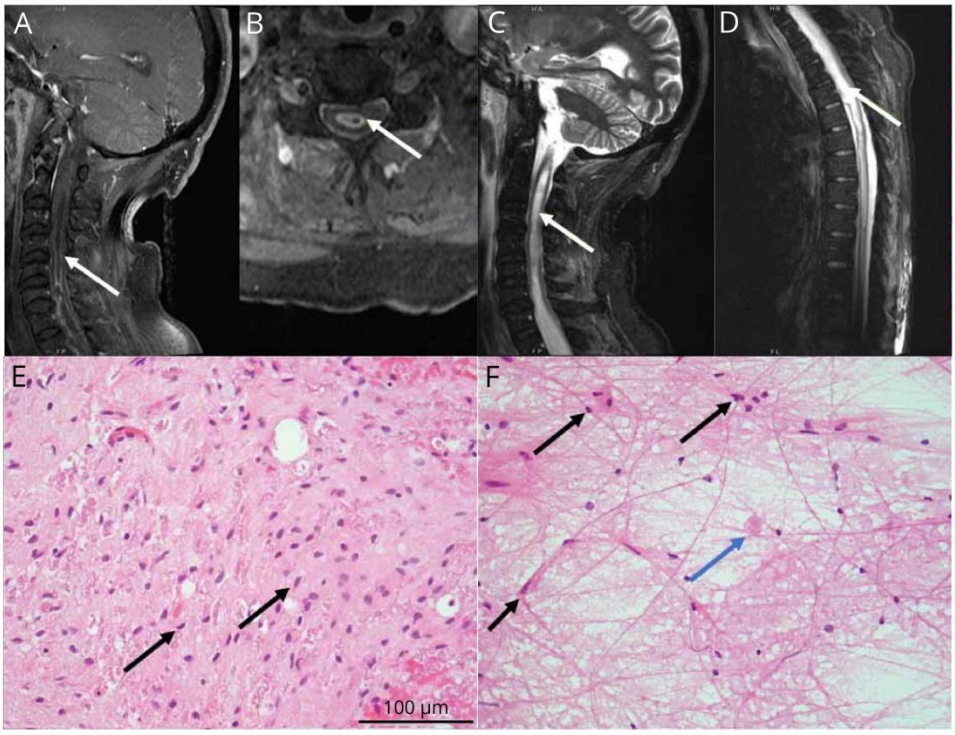
Figure: MRI of cervical and thoracic cord and H&E sections from thoracic cord biopsy
Spinal MRI reveals an expansile, intramedullary T2 hyperintense signal abnormality, with peripheral nodular enhancement spanning C4-T5 (A,B), and longitudinally extensive expansion of the central canal from obex to T11 **(**C,D**)**. H&E sections at 400x magnification show infiltrating glioma with moderately pleomorphic, hyperchromatic cells with piloid processes (E,F black arrows) and occasional eosinophilic granular bodies (F, blue arrow).



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