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

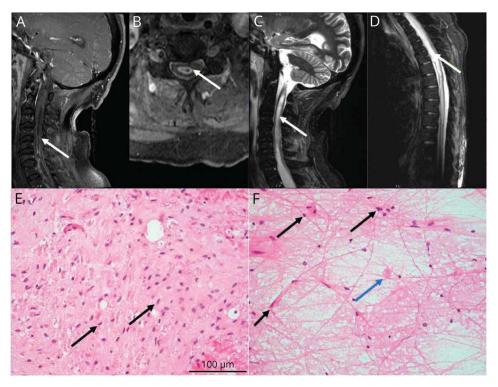
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Figure MRI of Cervical and Thoracic Cord and H&E Sections From Thoracic Cord Biopsy



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 400× magnification show infiltrating glioma with moderately pleomorphic, hyperchromatic cells with piloid processes (E, F black arrows) and occasional eosinophilic granular bodies (F, blue arrow). H&E = hematoxylin and eosin.

A 68-year-old man without a medical history developed 2 months of progressive weakness and cervicalgia. Examination 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 diagnostics were normal (eTable 1, links.lww.com/WNL/C653). CSF showed protein 2,505 mg/dL, 0 cells/ μ L, glucose 88 mg/dL, and CSF cell-free DNA sequencing identified a pathogenic variant in TERT p.C250T, suspicious for glioma. Thoracic spinal cord biopsy was pursued to exhaust reversible etiologies and revealed infiltrating glioma with TERT promoter mutation (Figure, E and F). Owing to progressive quadriplegia, respiratory failure, and poor prognosis, care was directed toward comfort.

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Spinal masses are classified as extradural, intradural extramedullary, or intradural intramedullary. Differential diagnosis for intramedullary cord lesions includes demyelination, paraneoplastic myelopathies (e.g., anti-CRMP5), neurosarcoidosis, infection, vascular abnormalities (e.g., dural arteriovenous fistula/malformation), nutritional deficiency, toxic insult, or tumor. Although noninvasive 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.

Author Contributions

S. Gritsch: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. Y. Aghajan: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. L. Kozanno: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. D. Chiu: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. J. T. Jordan: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. M. P. Frosch: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. G. Shankar: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data. W. Taylor Kimberly: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data.

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