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

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Figure Count: 1

Table Count: 0

Search Terms:

Acknowledgment:

Study Funding:
The authors report no targeted funding
Disclosures:
S. Gritsch reports no disclosures relevant to the manuscript; Y. Aghajan reports no disclosures relevant to the manuscript; L. Kozanno reports no disclosures relevant to the manuscript; D. Chiu reports no disclosures relevant to the manuscript; J.T. Jordan reports consulting income from Navio Theragnostics, Recursion Pharmaceuticals, CEC Oncology, holds stock in Navio Theragnostics and The Doctor Lounge and receives royalties from Elsevier; M. P. Frosch reports no disclosures relevant to the manuscript; G. Shankar reports no disclosures relevant to the manuscript; W. T. Kimberly reports consulting fees from NControl Therapeutics, research grants from Biogen, and equity in Woolsey Pharmaceuticals.

Preprint DOI:

Received Date:
2022-10-05

Accepted Date:
2023-01-19

Handling Editor Statement:
Submitted and externally peer reviewed. The handling editor was Resident and Fellow Section Editor Whitley Aamodt, MD, MPH.
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. 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. 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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References


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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Simon Gritsch, Yasmin Aghajan, Liana Kozanno, et al.
Neurology published online February 20, 2023
DOI 10.1212/WNL.0000000000207148

This information is current as of February 20, 2023

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