

Profound Leptomeningeal and Intramedullary Abnormalities in Bing-Neel Syndrome

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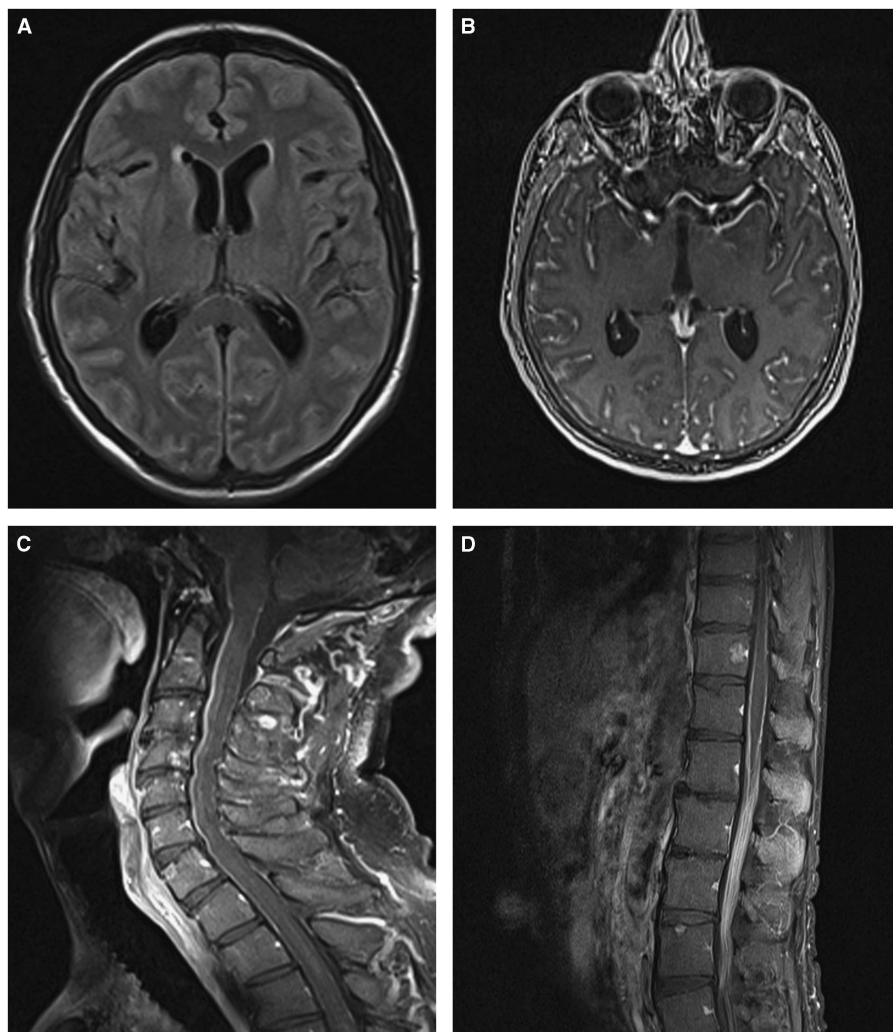


Figure 1: (A) Diffuse sulcal hyperintensities on FLAIR. (B) After gadolinium administration, there is extensive leptomeningeal enhancement in the brain, (C) cervical spine and (D) cauda equina.

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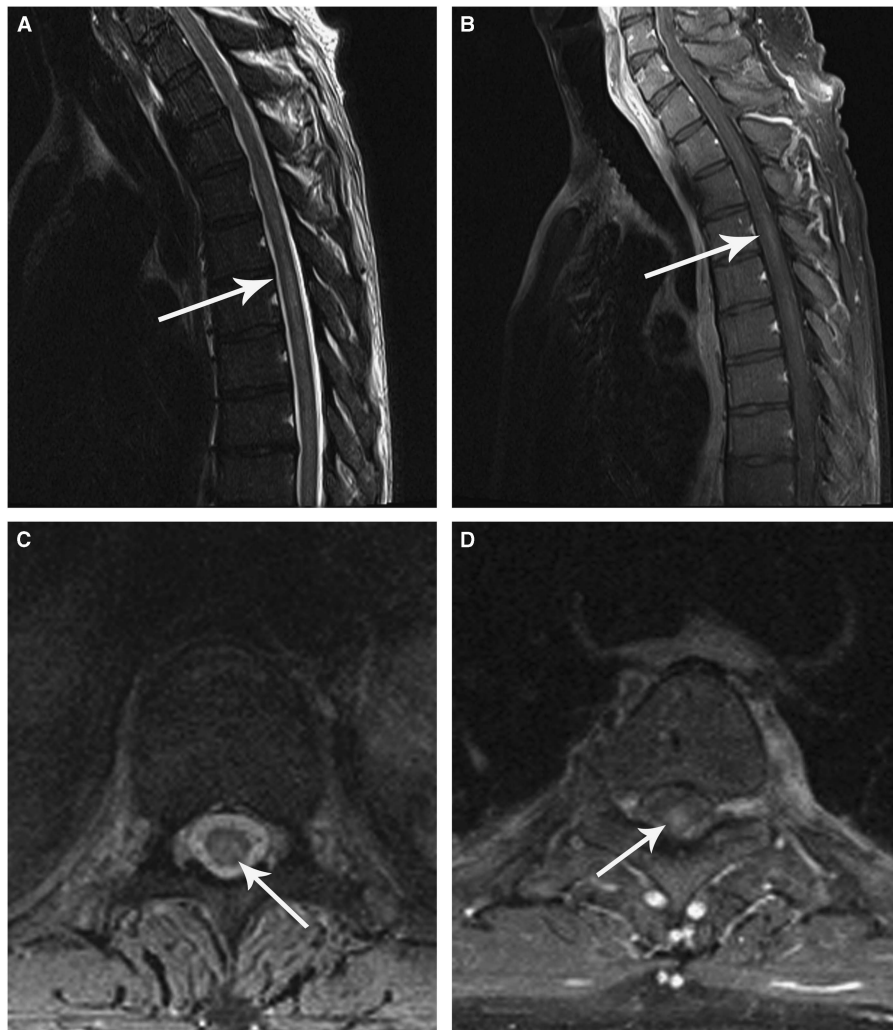


Figure 2: (A, C) MRI of the spine showing longitudinally extensive intramedullary hyperintensity on T2. (B, D) This lesion enhances with gadolinium.

A 52-year-old man with known Waldenström macroglobulinemia (WM) presented with acute psychosis and hallucinations. His neurologic examination was intact aside from abnormal mental status. He was initially admitted to psychiatry department for management of psychosis and hallucinations. Neurology was consulted to rule out organic cause of his persistent psychosis.

MRI showed extensive leptomeningeal enhancement of the entire brain, spinal cord and cauda equina (Figure 1). In addition, enhancing longitudinally extensive intramedullary T2 hyperintensity in the thoracic cord was noted (Figure 2). CSF showed markedly elevated protein (6.5 g/L) and lymphocytic pleocytosis, and flow cytometry was positive for central nervous system (CNS) lymphoplasmacytic lymphoma consistent with Bing-Neel syndrome.

Bing-Neel syndrome is a rare presentation of WM. Waldenström macroglobulinemia is a rare B-cell-mediated immunoglobulin M-producing lymphoproliferative disorder accounting for 1-2% of non-Hodgkin's lymphoma.¹ Bing-Neel syndrome is caused by infiltration of malignant B cells into the CNS, sometimes in the absence of systemic progression.² Patients present with progressive symptoms over weeks or months, including

headache, seizures, cognitive changes or cranial neuropathies from leptomeningeal involvement.² They can also present with hyperviscosity syndrome leading to visual loss (retinal hemorrhage) or epistaxis. Treatment is similar to primary CNS lymphoma, with steroid therapy as induction, followed by blood-brain barrier penetrating chemotherapy (e.g., methotrexate, cytarabine and ibrutinib).² Median survival for WM is estimated to be 7-12 years.² The survival rate for Bing-Neel syndrome is reported as 71% at 5 years and 59% at 10 years.³

Clinicians should remember that malignant B-cell infiltration to the CNS may lead to isolated psychiatric symptoms and should have low threshold for work-up of new psychosis or hallucinations in WM patients. In addition to leptomeningeal and intraparenchymal brain lesions, intramedullary spinal lesions may also be seen in Bing-Neel syndrome.

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DISCLOSURES

JZD, RG and SC have nothing to disclose.

STATEMENT OF AUTHORSHIP

JZD contributed to clinical care of the patient, case concept and drafting and revising the manuscript. RG was involved in drafting and revising the manuscript. SC was involved in imaging acquisition and drafting and revising the manuscript.

REFERENCES

1. Castillo JJ, D'Sa S, Lunn MP, et al. Central nervous system involvement by Waldenström macroglobulinaemia (Bing-Neel syndrome): a multi-institutional retrospective study. *Br J Haematol*. 2016;172(5):709-15.
2. Minnema MC, Kimby E, D'Sa S, et al. Guideline for the diagnosis, treatment and response criteria for Bing-Neel syndrome. *Haematologica* [Internet]. 2017;102(1):43-51.
3. Simon L, Fitsiori A, Lemal R, et al. Bing-Neel syndrome, a rare complication of Waldenström macroglobulinemia: analysis of 44 cases and review of the literature. A study on behalf of the French Innovative Leukemia Organization (FILO). *Haematologica*. 2015;100(12):1587-94.