Teaching NeuroImage: Bing-Neel Syndrome Mimicking a Meningioma With a Frontal Subcutaneous Lesion

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Rong Ge: Drafting/revision of the manuscript for content, including medical writing for content;
Study concept or design

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A 66-year-old woman presented with dizziness and anemia. Brain MRI revealed an intradural lesion with adjacent vasogenic edema in the right frontal and parietal lobes with dural tail sign and an arc-shaped homogenously enhancing mass in the subcutaneous soft tissue of the frontal region (Figure 1). Differential diagnosis for this lesion included meningioma, glioma, CNS lymphoma, or autoimmune disorders. Total resection of both lesions revealed pathological morphology consistent with lymphoplasmacytic lymphoma (Figure 2). Detection of an L265P mutation in the MYD88 gene in both CSF and bone marrow biopsy confirmed Bing-Neel syndrome, a rare neurological manifestation of Waldenstrom Macroglobulinemia (WM)\textsuperscript{1}. The MYD88 gene encodes a vital protein involved in immune system signaling, with 95\% of WM patients exhibiting an L265P mutation. The most common imaging findings are leptomeningeal infiltration or parenchymal involvement of the brain\textsuperscript{2}. Hematological malignancy should be considered when a meningeal lesion is accompanied by an extracranial tumor.
Figure 1 MRI Images of the Brain
(A) T1 and (B) T2-weighted axial images demonstrate a broad basal lesion (arrowheads) with vasogenic edema. Sagittal (C) and coronal (D) postcontrast T1 Flair images demonstrate an enhanced intradural lesion with dural tail sign (arrows), and an arc-shaped homogenously enhancing mass in the subcutaneous soft tissue (dotted arrows).
Figure 2 Pathology Specimen
Hematoxylin-eosin staining at 40X (A) and 200X (B) show diffuse dural involvement (yellow arrow, A) and perivascular infiltration (red arrows, B) by lymphocytes with plasmacytoid features. (C) 200X Immunohistochemical staining shows lymphocytes are positive for CD20 (red arrows).
References
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