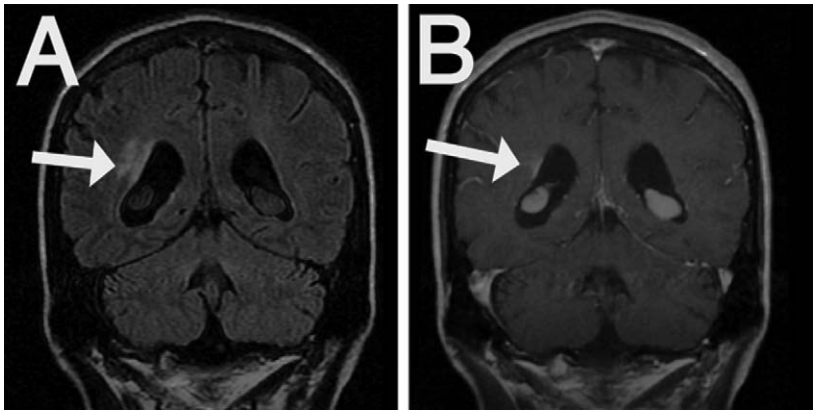
Imaging of Bing-Neel syndrome

Figure Brain MRI revealing a T2 prolongation (A: coronal FLAIR, arrow) in the right parieto-occipital periventricular white matter with patchy enhancement (B: post-gadolinium coronal T1, arrow)



A 64-year-old man with IgM kappa restricted Waldenstrom macroglobulinemia (WM) presented with right frontal headache and transient expressive aphasia. MRI demonstrated regions of enhancement within periventricular white matter with associated FLAIR hyperintensity and no restricted diffusion (figure). CSF analysis showed leukocytes 2 cells/mm^3 (71% lymphocytes), erythrocytes 73 cells/mm^3, elevated protein of 436.4 mg/dL, glucose of 48 mg/dL, and an M-spike of 34.5 mg/dL (IgM kappa restricted). CSF cytology showed a population of lymphoplasmacytoid cells and occasional plasma cells. This constellation of findings is suggestive of CNS involvement of WM (Bing-Neel syndrome) and biopsy was not performed. The patient is presently undergoing treatment with a CNS penetrating chemotherapy agent (temozolomide). Neurologic symptoms in patients with WM should raise suspicion of Bing-Neel syndrome, a rare complication of WM characterized by lymphoplasmacytoid infiltration of the brain parenchyma and IgM deposition in the CNS.[1] In patients with WM and neurologic presentation, CSF analysis and, if inconclusive, biopsy should be considered to rule out alternative neoplastic or infectious processes.

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