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Bing-Neel Syndrome: A Neuropsychological Case Study
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Objective: Central nervous system involvement by malignant cells is a rare complication of Waldenstrom macroglobulinemia (WM), known as Bing-Neel syndrome (BNS). BNS is even rarer than WM and involves central nervous system (CNS) infiltration by neoplastic lymphoplasmacytoid and plasma cells with or without cerebrospinal fluid (CSF) hyperglobulinemia. Due to the rarity of this diagnosis, few descriptions of neuropsychological performance exist in the literature. This case study highlights the neurobehavioral characteristics WM with confirmed BNS. We aim to expand understanding of cognitive effects of this rare diagnosis.

Method: A 71 year old Caucasian male physician underwent brain MRI, lumbar puncture (LP), and serial neuropsychological testing. Initial symptoms included disorientation, anomia, motor difficulties, and short-term memory disturbance. Due to initial neuropsychological test results in the context of WM, BNS was suspected and confirmed by LP.

Results: Initial neuropsychological testing showed severe problems with executive functioning and mild deficits in dexterity, as well as relative problems with verbal and visual fluency, processing speed, and visuomotor scanning. Fludarabine was used to treat the patient’s BNS with good success based on imaging and LP. Gait, mental status, and visual spatial difficulties improved. Despite treatment, the patient’s neuropsychological profile showed declines in memory, fluency, attention, processing speed, dexterity, and executive functioning.

Conclusion: This case study highlights the neuropsychological findings in a rare complication of WM. As this case demonstrates, negative LP and MRI do not completely rule out BNS. Other treatments that cross the blood brain barrier may need to be considered to fully treat the CNS complications of BNS. A comprehensive approach, that includes neuropsychological evaluation, is recommended to elucidate cognitive deficits in WM.