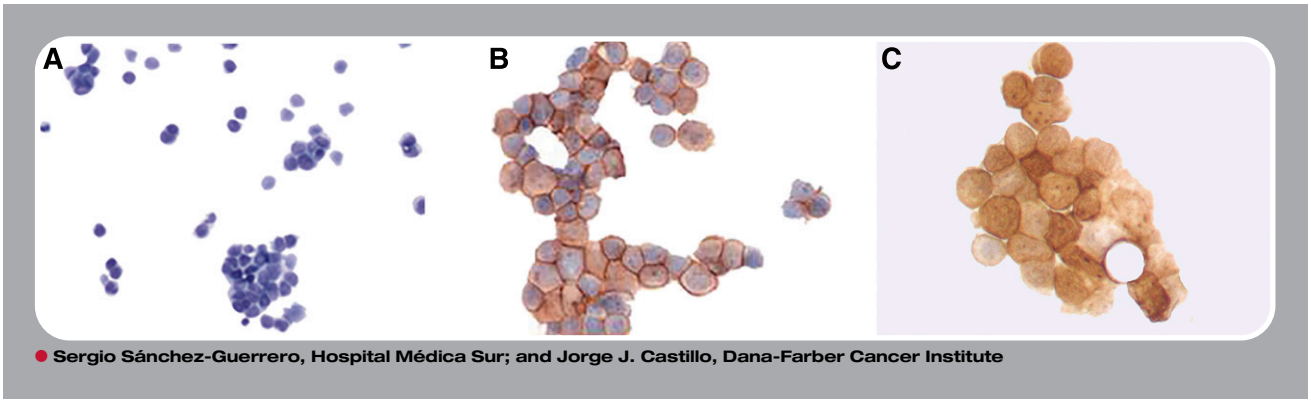
bloodwork
IMAGES IN HEMATOLOGY

Bing-Neel syndrome: a rare complication of Waldenström macroglobulinemia



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A 61-year-old woman with Waldenström macroglobulinemia (WM) was treated with 6 cycles of cyclophosphamide, rituximab, and dexamethasone between December 2011 and May 2012, and she achieved complete remission. She was given maintenance rituximab once every 3 months. Eighteen months later, the patient presented with complaints of headache and blurry vision. Physical examination revealed aphasia, paraparesis, and nuchal rigidity. A magnetic resonance imaging scan of the brain showed leptomeningeal enhancement without signs of encephalitis, stroke, or tumor. Cerebrospinal fluid (CSF) showed leukocytes 64/μL (95% mononuclear cells), glucose 9.8 mg/dL, and protein 110 mg/dL. CSF cytology and immunohistochemistry showed κ-restricted B lymphocytes. The figure shows Waldenström cells in CSF with Wright staining (panel A), CD20 expression (panel B), and kappa light chain restriction (panel C). A diagnosis of Bing-Neel syndrome (BNS) was made. The patient was treated with 6 doses of intrathecal (IT) methotrexate and hydrocortisone twice a week with reduction of neoplastic cells in the CSF. Four months later she relapsed, and an allele-specific polymerase chain reaction for $MYD88^{L265P}$ in CSF was positive. Intravenous fludarabine was initiated with plans to add IT liposomal cytarabine, but she refused further treatment and died at home 7 months after the diagnosis of BNS.

First reported in 1936, BNS should be suspected in patients with WM and neurologic impairment. There is no standard of care for BNS, and prospective studies are needed to address the optimal treatment of this late, rare, and dangerous complication of WM.

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