bloodwork
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Crystal-storing histiocytosis in Bing-Neel syndrome

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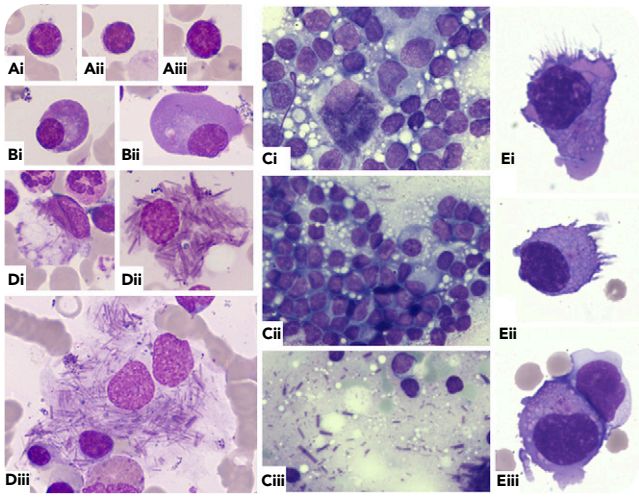

A 69-year-old man was admitted for abdominal pain and confusion. He presented with lymphadenopathy, hepatosplenomegaly, and polyneuropathy. Brain magnetic resonance imaging scans revealed white matter lesions. An immunoglobulin-M λ monoclonal protein was also detected (4.2 g/L). Bone marrow (BM) aspirate showed involvement with 15% of the differential lymphoplasmacytic cells (panel Ai-iii, original magnification ×100, May-Grunwald-Giemsa [MGG] stain) and plasma cells with pink cytoplasmic inclusions (panel Bi-ii, original magnification ×100, MGG stain), suggesting the diagnosis of lymphoplasmacytic lymphoma/Waldenstrom macroglobulinemia. This was confirmed by BM biopsy (nodular lymphoplasmacytic/plasma cell involvement) and lymph node (LN) biopsy (panel Ci-iii, imprints from biopsy, original magnification ×40, MGG stain). Flow cytometry of the BM sample detected λ monotypic B cells and plasma cells. The MYD88 L265P mutation was not found by next-generation sequencing on BM and LN samples. However, rare histiocytes with crystalline cytoplasmic inclusions were identified on the BM smears (panel Di-iii, original magnification ×100, MGG stain), LN imprints (panel Ci, original magnification ×40) and colonic biopsy, supporting crystal-storing histiocytosis, which is a rare lesion composed of histiocytes with abnormal intra-lysosomal accumulation of immunoglobulin reported mainly in patients with plasmacytic/lymphoplasmacytic neoplasms, but also in inflammatory conditions. Interestingly, lymphoplasmacytic and plasma cells with pink inclusions (panel Ei-iii, original magnification ×100, MGG stain) also infiltrated the cerebrospinal fluid. The patient was treated with *R*-bendamustine and is still alive after 1 year of treatment.

This case illustrates a crystal-storing histiocytosis associated with an underlying Bing-Neel syndrome.

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