EPIDURAL METASTASIS AS PRESENTATION OF BING-NEEL SYNDROME: A CASE REPORT
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Introduction
Waldenstrom macroglobulinemia (WM) is a lymphoplasmocytic lymphoma with bone marrow and lymphatic tissue involvement and IgM monoclonal gammopathy. Neurological complications occur in approximately 25% of patients. Malignant infiltration of the central nervous system (CNS) or leptomeningeal spaces is very rare and referred to as Bing-Neel syndrome (BNS). Epidural metastasis causing spinal cord compression has been reported exceptionally in WM.

Case report
In 2005 a 63 year-old man was diagnosed with WM. In 2014, he was referred to our neurological department due to a 2 month history of difficulty in walking, stiffness in the legs, hand clumsiness and episodic urinary urgency. Serum protein electrophoresis and immunofixation confirmed the monoclonal IgM kappa light chains. Cerebrospinal fluid (CSF) examination revealed 96 cells/mm³ with 90% of lymphocytes, low glucose concentration and increased protein level (1288 mg/dl). CSF cytology and flow cytometry were negative for malignant cells. Brain magnetic resonance imaging (MRI) showed leptomeningeal enhancement at the brainstem level. Cervical and dorsal MRI disclosed an epidural mass spanning between C2 and C4 levels, with intense gadolinium enhancement, causing spinal cord compression and hyperintensity at T2-weighted sequences, and diffuse leptomeningeal enhancement (fig. 1A/B). The patient initially received glucocorticoids with partial improvement of the clinical picture. Thereafter, he underwent a decompressive laminectomy with complete excision of the epidural mass. Pathological examination revealed a diffuse lymphoid infiltrate of small lymphocytes, plasmacytoid lymphocytes, and plasma cells. (fig 1C/D). Immunohistochemical staining of these cells was positive for CD20, CD79 and partially for CD38. Staining for immunoglobulin light chains revealed kappa light chain. These results supported the diagnosis of epidural localization of WM. The patient underwent systemic combined chemotherapy followed by autologous hematopoietic cell transplantation. At 3-month follow-up, the patient reported a recovery of hand clumsiness and bladder urgency, and an improvement of walking that was unaided.

Conclusion
Neurological complications occur in about 25% of patients with WM. Direct infiltration of malignant cells into the CNS is a rare complication known as BNS, with only 32 cases reported in literature. Spinal cord involvement in WM is considerably less frequent than brain involvement although isolated cases of myelopathy, cauda equina amylosis and extrinsic compression by vertebral osteolysis have been reported.

References