Here, we describe a 52-year-old woman with a history of primary plasma cell leukemia (PCL) who achieved complete remission after the tandem of autologous and nonmyeloablative allogeneic stem cell transplantation (SCT). Nine months after the allogeneic SCT, she presented with oculomotor and trochlear nerve palsy (diplopia, ptosis of the right upper eye lid, and dilated right pupil). Physical examination also revealed mild paresis of the upper arm muscles and the muscles involved in dorsiflexion of the left foot. Contrast-enhanced craniospinal magnetic resonance imaging showed multiple enhancing leptomeningeal nodules (panel A arrows). Cerebrospinal fluid (CSF) examination revealed 321 × 10^6 leukocytes per L and an elevated total protein level of 2.08 g/L. Microscopic analysis of the CSF showed the presence of very atypical plasma cells (panel B), some of which contain intranuclear inclusions (Dutcher bodies). Immunophenotypic analysis confirmed the presence of plasma cells in the CSF (60% of all nucleated cells). These cells had an aberrant phenotype (CD38^− CD138^− CD19^− CD56^− CD45^− ); panel C). There was no systemic activity of the primary PCL.

Primary PCL more frequently presents with extramedullary disease when compared with classic multiple myeloma. Therefore, in cases involving neurological deficits, leptomeningeal tumor infiltration should be considered both at diagnosis and at relapse. CSF investigation is mandatory at an early stage to prevent a delay in diagnosis and treatment.
Leptomeningeal metastases from primary plasma cell leukemia

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