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Case Report

Waldenström's macroglobulinemia and cerebral lymphoplasmocytic proliferation: Bing-Neel syndrome

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Waldenström's macroglobulinemia (WM), a lymphoplasmacytic lymphoma, is characterized by neoplastic proliferation of B lymphocytes and excessive production of monoclonal immunoglobulin M (IgM). The leading clinical features are lymphoadenopathy and/or splenomegaly, anemia and hyperviscosity syndrome. Cognitive impair-ment due to central nervous system infiltration by lymphoplasmocytoid cells (Bing-Neel syndrome) has been rarely reported (Civit et al., 1997).

A 65-year-old Moroccan woman was admitted to our department because of headache, dizziness, progressive muscle weakness, and diffuse bone pain. There was no sign of lymphadenopathy, nor liver or spleen enlargement on clinical examination. Hematologic laboratory values were as follows: haemoglobin 11.8 g/dl; white cell count 4.1x109/L with a normal differential count and platelets 170x109/L. Blood chemistry and urine tests were normal. Serum protein immunoelectrophoresis revealed the pre-sence of 8.4 g/dl monoclonal IgM-kappa gammopathy. Bone marrow biopsy showed hypercellularity with inter-stitial and paratrabecular aggregates of plasma cells and lymphocytes, which were mostly small and round-shaped. Bone marrow flow cytométrie disclosed a popu-lation of kappa-positive lymphoid cells expressing CD5, CD19, CD22 and CD38. A diagnosis of lymphoplasma-cytic lymphoma was made and 3 courses of CVP (cyclo-phosphamide 300 mg/m^2 per day on days 1-5 p.o, vin-cristine 1.4 mg/m^2 on day 1 i.v, and prednisone 100 mg per day on days 1-5 p.o.) were administered. Clinical symptoms improved and the patient was discharged. Four months after last course the patient was readmitted to hospital because of persistent headache and increased IgM levels. A brain computed tomography scan revealed multifocal extra-axial tumorous lesions along the dura matter. A brain magnetic resonance imaging showed an extra-axial soft tissue tumor along the left cavernous sinus and tentorium, right frontal convexity and tentorium and falx; the brain parenchyma appeared to be unremarkable. Cerebrospinal fluid analysis showed the following: white cell count 39/ L with neutrophils 2%, lymphocytes 51% and monocytes 47%, total protein 1.71 g/dL, glucose 108 mg/dL, IgM 67.5 mg/dL and a few plas-macytoid lymphocytes on cytology. Stereotactic biopsy of dural tissue at the falx showed a diffuse infiltration with atypical cells, which were identified immunophenoty-pically as plasmacytoid lymphocyte. The patient was con-firmed to have CNS infiltration by atypical plasmacytoid lymphocyte infiltration (Bing-Neel syndrome). The patient refused irradiation therapy and died of multiple organ fai-lure within a few days of admission to hospital.

In 1936 Bing and Neel reported the association of hyperglobulinemia, CNS symptoms (paresthesias, headache, and paralysis), and brain infiltration composed of plasma cells and lymphocytes in two patients (Bing and Neel, 1936). The Bing-Neel syndrome appears to be the result of involvement of the CNS by diffuse neoplasm infiltration. Neurological complications occur in about 25% of patients with WM. Although they are most often peripheral, they can involve the CNS. In WM, the CNS may be involved by a variety of mechanisms, including hyperviscosity and direct infiltration by neoplastic cells (Scheithauer et al., 1984). Patients with Bing-Neel syn-drome have sometimes presented with a mass containing neoplastic cells, but the masses have been intraparen-chymal rather than meningeal (Imai et al., 1995).

Histological confirmation is necessary to establish the definitive diagnosis. The outcome for most patients who underwent chemotherapy was poor, and the patients died within several months.

Recently, a great deal of interest has been noted by treatments with purine nucleoside analogs (fludarabine, cladribine, and pentostatin) because of their remarkable activity in lymphoproliferative disorders. It has been reported that a patient with Bing-Neel syndrome (in its diffuse form) was successfully treated with cladribine administration, or radiation therapy and combination of cladribine, cyclophosphamide, and prednisone (Delgado et al., 2002). Several retrospective and prospective stu-

dies have indicated that rituximab may induce an objective response in approximately 30 - 40% of previous treated patients with WM. However, the effect of rituximab treatment on the cerebrospinal fluid B- cell compartment is limited in comparison with the effect on the B cells in the periphery (Monson et al., 2005), and it has not been tried in Bing-Neel syndrome as yet. Therefore, the effect of rituximab on CNS involvement of WM needs to be validated by of future studies.

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