Ictus cerebellopontine as first manifestation of an intravascular diffuse large B cell lymphoma involving at the kidney

Dear Editor:

Intravascular diffuse large B-cell lymphoma (IVDLBCL) is a rare variant of the extranodal diffuse large B-cell lymphoma characterized by a selective growth of lymphomatous cells at the blood vessels lumen area, especially in capillaries, avoiding large vessels. This type of lymphoma is mainly diagnosed in adults of approximately 67 years, on average, and is not predominant between genders. There are 2 main clinical cases: Western cases, where damages are predominantly neurological and cutaneous, and Asian cases, which present multiple organ failure, pancytopenia, and hemophagocytic syndrome. This type of lymphoma has a limited response to chemotherapy, and its prognosis is poor.

We present the case of a woman, 66 years, with no allergy to medications and no toxic habits, who was admitted to the Neurology Department of the Hospital Sant Joan Despí Moisès Broggi hospital (HSJDMB) in October, 2012, with an acute cerebellopontine ischemic infarction, diagnosed by MRI, and who recovered from it ad integrum. During her hospitalisation, she had episodes of fever.
not predominantly in the evening, with a series of negative hemocultures. The presence of bicytopenia stood out in the analysis, with Hb 100 g/l, a platelet count of 70 x 10^9/l, an ESR of 120 mm/first hour, LDH 480 U/l, and Beta-2 microglobulin 4.25 mg/dl; kidney and liver functions were preserved, and the 24-hour urinalysis was strictly normal. The patient underwent a CT scan, showing homogeneously hepatosplenomegaly and a bilateral renal mass. She was transferred to the HSGMB Internal Medicine Unit for the study of the mass, and an ultrasound-guided renal biopsy was performed. The anatomopathological results obtained were indicative of a diagnosis of an IVDLBCL, affecting glomerular bone vessels lumen. The immunohistochemistry was positive for CD20 and for negative for BCL2 and BCL6; Ki67 was >70% and showed a clonal ordering of immunoglobulin heavy chain. A PET was done for additional testing, showing pathological spreading affects hepatosplenic structures, kidneys, both suprarenal glands, uterus and bone structures (left iliac crest, and vertebrae C7, T10–T12 and L2–L5). The biopsy of the bone marrow showed a sinusoidal infiltration caused by lymphoma, with preservation of the haematopoiesis. A lumbar puncture was performed, indicating infiltration by lymphoma.

The definitive diagnosis was stage IV B IVDLBCL by infiltration of bone marrow, CNS, hepatosplenic, renal, suprarenal, uterine and multiple-bone structures, IPI 5/5. The patient received a first-line treatment following the R-CHOP (rituximab, cyclophosphamide, adriamycin, vincristine and prednisone) protocol, as well as a triple intrathecal chemotherapy treatment consisting of methotrexate cytarabine and hydrocortisone, for a total of 8 cycles, obtaining complete remission (CR). In May 2013, consolidation therapy was given to the patient through peripheral blood hematopoietic progenitors autologous transplant, with a preparation according to the scheme BEAM (BCNU, Etoposide, Ara-C and Melphalan). The patient, who was re-evaluated by PET, general analysis, and medullar biopsy, has remained in CR 5 months after the transplant.

IVDLBCL is a rare variant of the diffuse large B-cell lymphoma in Western clinical cases, most frequently affecting the CNS and cutaneous structures, and presenting fever without a focus.1,4–6

The first case of IVDLBCL diagnosed by means of a renal biopsy was described in 1981,7 and has been reviewed 21 times since then.6,8,9 This lymphoma is most commonly located in the renal glomerular capillary lumen area.5 Some of the most frequent clinical findings in cases of IVDLBCL affecting kidneys are acute renal failure with urine protein in nephrotic range,9 as opposed to the case we described above.

Since there is no standard treatment for IVDLBCL, chemotherapy treatment schemes for aggressive types of lymphomas are administered, preserving the CNS, since it is frequently damaged. Response to chemotherapy is poor,1 despite the fact that adding rituximab to standard treatment schemes has significantly improved the management of this type of lymphoma, with an increase in progression-free survival rates and global survival rates10.

Lastly, neurological disturbances, as well as the presence of pancytopenia, elevated levels of LDH and Beta-2 microglobulin, with or without multiple organ failure, should lead to suspect the presence of an underlying lymphoproliferative disorder similar to the one presented by the abovementioned female patient.1,2

References


Miguel Sagüés a,b,*, Viviana Paredes a, Jordi Altés b, Alberto Fernández de Sevilla a

a Servei d’Hematologia Clínica, Institut Català d’Oncologia-Hospital Duran i Reynals, Universitat de Barcelona, L’Hospital de Llobregat, Barcelona, Spain
b Servei de Medicina Interna, Consorci Sanitari Integral-Hospital Sant Joan Despí Moisès Broggi, Sant Joan Despí, Barcelona, Spain

* Corresponding author.

E-mail address: michel.sagues.serrano@gmail.com (M. Sagüés).