Angiotropic lymphoma: a rare hematological malignancy

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A 56-year-old woman was referred to us complaining of painful subcutaneous nodules in the lower extremities during the last three months. The nodules appeared a year ago. The physical examination disclosed palpable nodules in the medial surfaces of the thighs, each with a maximal diameter of 1.5 cm. The diagnosis was made after the biopsy of a nodule from the thigh (Figure 1). The skin and the subcutaneous tissue presented dilated small vascular ramifications, whose lumens were filled with aggregates of large, atypical lymphoid cells. These lymphoid cells were LCA$^+$ and L26$^+$, and there was no monoclonal κ or λ chain expression; also, in order to exclude T-cell lineage, CD3 and CD45RO antigens were assayed and found to be negative.

The patient was treated with CHOP-bleomycin (8 cycles) plus methotrexate intrathecally (5 cycles). After the initial three cycles of CHOP-bleomycin the physical examination was negative for nodules and CT scans of the head, chest and abdomen showed no evidence of disease. The disease relapsed after the completion of the seventh chemotherapy cycle as shown in a new nodule biopsy, this time with bone marrow involvement. The patient received a single cycle of ESAP with no response and died following complications of septic shock.

Angiotropic lymphomas (also referred to as intravascular malignant lymphomas, malignant endotheliomatosis or neoplastic angioendotheliosis) are rare and very aggressive intravascular non-Hodgkin's lymphomas of B-cell lineage.[1] Clinical findings include cutaneous lesions due to infiltration, progressive dementia, adrenal masses and fever of unknown origin.[2] Diagnosis is made at autopsy or by biopsy with the assistance of immunochemistry, ultrastructural analysis, cytogenetics or molecular biology studies.[3] Depending on the disease's localization, differential diagnosis includes panniculitis, thrombophlebitis, diffuse multisystem vasculitis, malignant neoplasms and vascular tumors;[3,4] also, angiocentric T-cell lymphomas can appear with subcutaneous nodules.[5] For practical reasons, angiotropic lymphomas are considered to be generalized at the time of initial diagnosis since central or peripheral nervous system involvement is found in at least half of the patients. In this rare condition, the involvement of bone marrow is more rare. Treatment of choice is CHOP or PROMACE, with less than half of the patients achieving complete remission.[2]

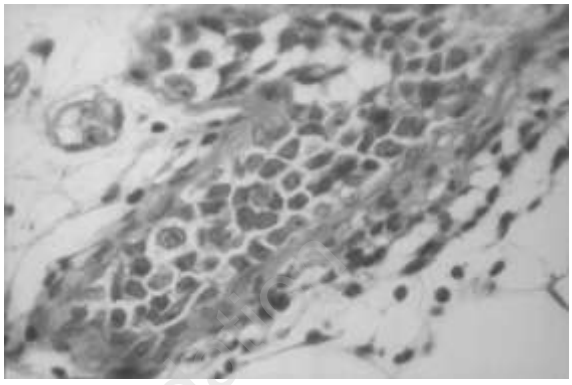

Figure 1. Vascular cross section showing intraluminal lymphoid cells aggregates (HE ×400).

References

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