

Diffuse Large B-cell Lymphoma, Leg Type A diagnosis not to forget in the elderly

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سرطان الغدد الليمفاوية ذو الخلايا B الكبيرة المنتشر تشخيص لا يجب نسيانه في كبار السن

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Figure 1: Infiltrated erythematous nodules on the anterior aspect of the left leg with severe hyperkeratosis and secondary impetiginisation.

AN 83-YEAR-OLD WOMAN, WITH A PAST medical history of hypertension, diabetes mellitus and ischemic cardiomyopathy, was referred to the dermatology clinic of Complejo Hospitalario de Jaen in Jaen, Spain, in October 2013. The patient presented with erythematous indurated tumoural lesions with severe hyperkeratosis and impetiginisation on the anterior aspect of her left leg, which had appeared eight months prior [Figure 1]. Fever, night sweats, weight loss, itching or other constitutional symptoms were absent. Neither hepatosplenomegaly nor axillary or generalised lymphadenopathy were detected. A systemic

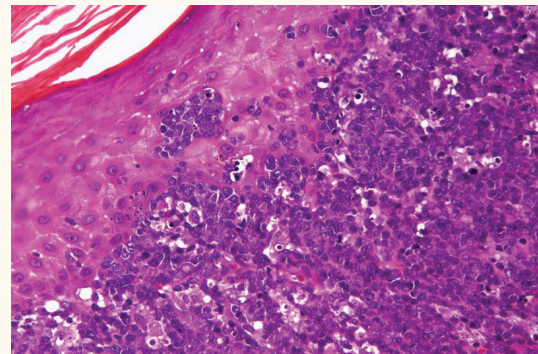


Figure 2: B-lymphocyte infiltration with a predominance of medium size and split non-cleaved cells with blastic differentiation.

examination of the patient did not reveal any further abnormalities.

All laboratory tests were negative, including a full blood count; biochemistry, protein electrophoresis, routine urine, lactate dehydrogenase, β 2-microglobulin and immunoglobulin tests; a peripheral blood smear, and serology tests for viruses (human immunodeficiency virus and hepatitis B and C viruses) and *Borrelia burgdorferi*. No evidence of malignancy was observed during a whole-body computed tomography scan or a bone marrow biopsy. A cutaneous biopsy was also performed.

A histological examination of the skin specimen revealed a B-lymphocyte proliferation with mild epidermotropism and antigen CD20/45 positivity. The following profile was obtained from an immunohistochemical assay: positive *multiple myeloma oncogene 1 (MUM-1)*, positive B-cell lymphoma 6 (Bcl-6), negative chromosome 10 (C10), positive B-cell lymphoma 2 (Bcl-2) and antigen Ki67 >90%. These findings were consistent with a diagnosis

of diffuse large B-cell lymphoma, leg type [Figure 2]. In addition, testing for the Epstein-Barr virus by *in situ* hybridisation was negative.

Comment

Diffuse large B-cell lymphoma, leg type is a subset of primary cutaneous lymphoma, characterised by the presence of nodules or tumours on the legs of elderly people, mainly females.¹ It represents 1–4% of all cutaneous lymphomas and has a five-year survival rate of less than 50%.² This form of lymphoma was identified as an independent entity in the classification of cutaneous lymphomas developed by the European Organisation for Research and Treatment of Cancer.³ Although these lymphomas are mostly limited to the skin at presentation, in 10% of reported cases the tumoural lesions begin in locations other than the legs.⁴ When the malignancy spreads, it frequently advances to the lymph nodes, bone marrow and the central nervous system.⁴

As the majority of the patients with this condition are elderly, the therapeutic management may be complicated due to comorbidities and the potentially frequent number of relapses. Spontaneous remission only occurs in rare cases.⁵ Rituximab plus cyclophosphamide, doxorubicin, vincristine and prednisolone (R-CHOP) chemotherapy and chemotherapeutic compounds applied for relapsing systemic B-cell non-Hodgkin's lymphoma have previously proven to be effective therapies.⁶ However, the current first line of treatment for this variety of lymphoma involves anti-CD20 antibodies, such as rituximab. These therapies can be used even during the early stages of the condition in order to improve rates of survival.⁷

References

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