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A PRIMARY CUTANEOUS LARGE B-CELL LYMPHOMA OF UNUSUAL LOCATION

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ABSTRACT

We report the case of a patient with a primary cutaneous large B-cell lymphoma (PCLBCL) of unusual location. A 65-year old man presented a left jugulo-mandibular infiltrated nodular placard rapidly evolving for two months with general deterioration. The cutaneous biopsy showed large cells infiltrate dissociating collagen fibers. Immunohistochemistry revealed a large B-cell lymphoma. Blood count, lymph node ultrasound, and thoraco-abdominal computed tomography were normal. The patient received six cycles of R-CHOP chemotherapy (Rituximab, Cyclophosphamide, Hydroxy Doxorubicine, Vincristine, Prednisone) with complete remission. PCLBCL of cephalic location have a better prognosis than that of the legs.

KEYWORDS: cutaneous B-cell lymphomas - primary cutaneous large B-cell lymphoma – Chemotherapy.

INTRODUCTION

Primary cutaneous B-cell lymphomas are rare tumors representing 25% of all primary cutaneous lymphomas.[1] Large cell histological type accounts for only 5%. Its location is often in the lower limbs, hence the term "leg type".[2] We report the case of a patient with a primary cutaneous large B-cell lymphoma of unusual location in the left mandibular region.

CASE REPORT

A 65 year old patient consulted for a left mandibular infiltrated and nodular placard evolving rapidly for two months. The history of the disease dated back two years with the appearance of a small retro-auricular nodule, which had rapidly increased in size over the last two months. The symptomatology evolved in a context of altered general condition with a weight loss of eight kilograms. Clinical examination found an erythematous, infiltrated, and ulcerated nodular placard with a fibrinous background, fifteen centimeters long and poorly limited in the left mandibular region, with no palpable adenopathy (Figure 1).

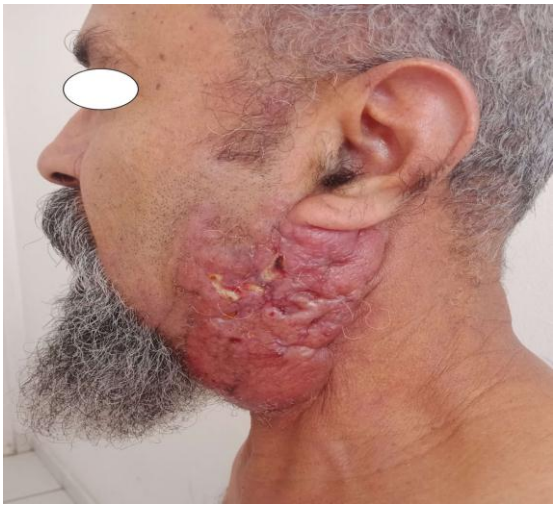

Figure 1: Infiltrated nodular placard in the left mandibular region.

The cutaneous biopsy showed a large cellular infiltrate dissociating collagen fibers, with ill-defined cytoplasm and round anisokaryotic nuclei (Figure 2). Immunohistochemistry was positive for CD20, CD3, CD10, BCL2, BCL6 and ki67, concluding to a large B-cell lymphoma (Figure 3).

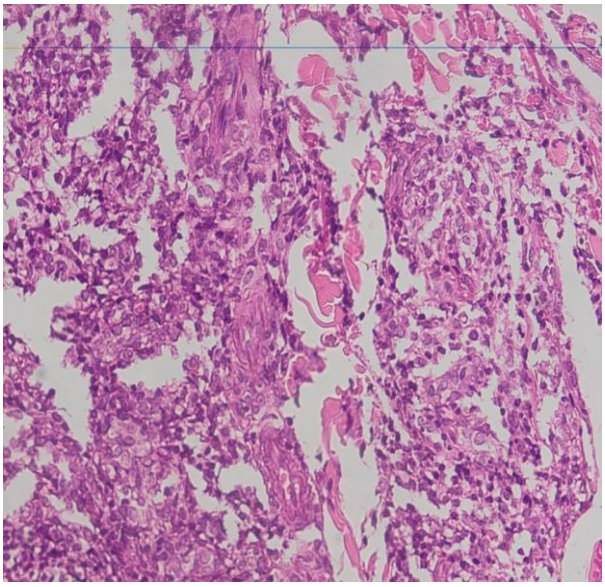

Figure 2: Histology showing large cellular infiltrates dissociating collagen fibers.

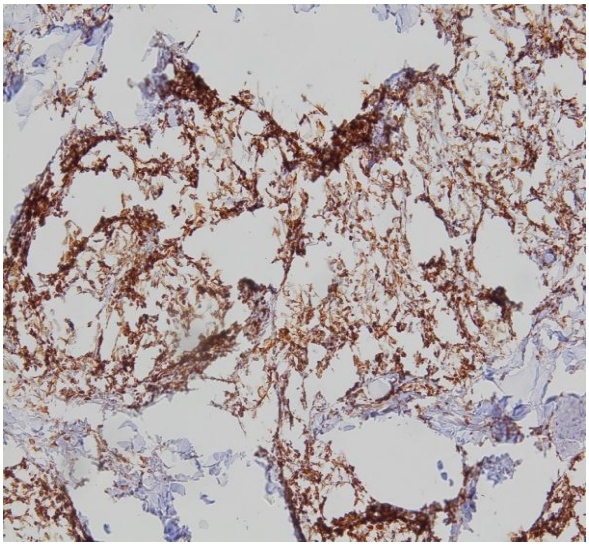

Figure 3: Positive Immunohistochemistry for BCL2.

Blood count, lactate dehydrogenases, lymph node ultrasound and thoraco-abdominal computed tomography were all normal. We retained the diagnosis of a primary cutaneous B-cell lymphoma. The patient received six cycles of R-CHOP chemotherapy with complete remission. The follow-up is two years without relapse or systemic involvement.

DISCUSSION

Primary cutaneous large B-cell lymphomas (PCLBCL) of cephalic location are rare accounting for only 0.56-5% of all PCLBCL and 2-18% of PCLBCL outside the lower limbs.[2] In a meta-analysis of 2021, only 7 cases of cephalic PCLBCL were reported. The locations were in order of frequency: leg, trunk, arm, and head.[3] PCBCL of cephalic location are characterized by their occurrence at a younger age than when localized in the legs (≈ 76 years), but also by the unique character of the lesions and the rarity of extra-cutaneous involvement.[4]

Risk factors of cutaneous B-cell lymphoma are Epstein Barr Virus, Lyme disease or immunosuppression.[5] Centro-follicular cutaneous B-cell lymphoma can sometimes rapidly progress to a large B-cell lymphoma.[6] In our patient, the presence of a retro-auricular nodule for two years and its rapid evolution during the last 2 months could be suggestive of this transformation, hence the interest to perform a cutaneous biopsy at the slightest doubt. The diagnosis is histologically confirmed with an immunohistochemical study. Large atypical lymphoid cells of immunoblastic or centroblastic morphology are found, of B phenotype in immunohistochemistry, labelled by BCL2 and MUM1.[7]

The treatment of cutaneous large B-cell lymphomas is based on a polychemotherapy R-CHOP (Cyclophosphamide, Doxorubicin, Vincristine, Prednisolone and Rituximab).[8] This is an aggressive treatment given their intermediate prognosis.[6] However, the prognosis of PCLBCLs located outside the lower limbs is better.[4]

CONCLUSION

We report a rare and unusually localized case of cutaneous primary large B cell lymphoma. It is true that the latter is of poor prognosis, but its location outside the lower limbs, especially cephalic, is generally of better prognosis.

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