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Diffuse large B cell lymphoma: cutaneous presentation

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DESCRIPTION

An 85-year-old patient was brought to the emergency department due to progressive weight loss and bed confinement over a period of 6 months. After recollecting a fully detailed medical history, the patient describes the growth of a gruelling mass on her right breast over the course of 1 year, with no mention of other concurrent symptoms such as fever or diaphoresis.

On her physical examination, there was an extremely emaciated patient with a performance status Eastern Cooperative Oncology Group (ECOG) 4, presenting with multiple subcutaneous nodules distributed mainly through her upper body, the largest in size being the one on her right breast already with evidence of tissue necrosis and coexisting right axillary lymphadenopathies (figure 1).

A full body CT Scan revealed uncountable lymph nodes mainly on the mediastinum, abdomen and pelvic regions, also with extensive subcutaneous and soft-tissue involvement of the right breast. For diagnostic purposes, the patient underwent a fine-needle biopsy of the right breast nodule, which histopathology revealed to be a large cell diffuse lymphoma non-germinal centre B cell CD20+, CD5+, MUM1+, bcl-2+, with a proliferation index of 70% compatible (figure 2).

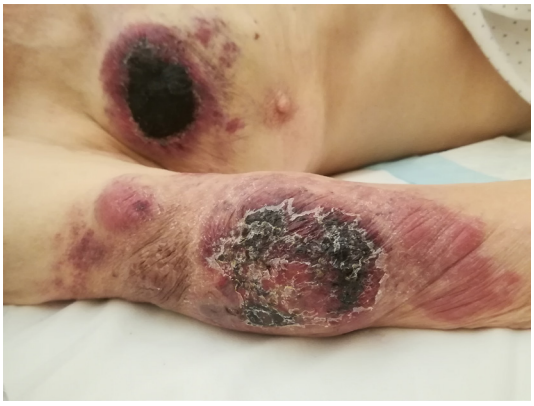

Figure 1 Large epicondylian mass with central area of necrosis and vasculitic alterations on the surrounding skin.

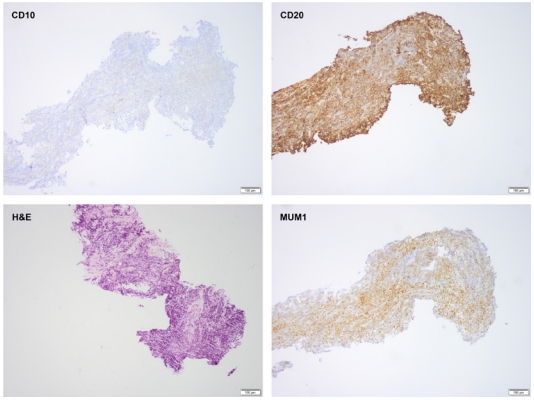

Figure 2 Needle biopsy affecting diffuse proliferation of medium to large atypical lymphocytes. Cell proliferation index of 70%. Immunohistochemical study reveal staining positive for CD20, MUM1 and negative for CD10—findings compatible with diffuse large B-cell lymphoma.

Learning points

- The differential diagnosis of primary nodular skin lesions opens a wide number of hypothesis making it a challenge for the clinician to investigate other signs and symptoms that allow for a better diagnostic approach.
- The first clinical sign of diffuse large B-cell lymphoma (DLBCL) is a quickly growing, non-painful mass, typically a lymph node in the neck, groin or abdomen, which may be accompanied with type B symptoms making this case an atypical presentation as for the initial absence of B symptoms but also the location of skin lesion.
- Subcutaneous nodule as the presenting sign of DLBCL is a rare form of primary presentation. The uncommon incidence of such manifestations in this subtype of neoplasia justifies reporting this case and highlights the importance of multidisciplinary teams in the management of patients with cancer.
- Onset presentation on an older female patient versus most current reports that focus on younger patients of the male sex with better outcomes at the point of diagnosis, makes this case explanatory of how diagnostic approach and prognostic indicators are relevant.
- The majority of cases reported of primary or secondary skin lesions of lymphoma focus on patients with singular, patchy or smaller dimension nodules making this presentation less frequent.
- This case is explanatory of how an adequate evaluation of patient's performance status and burden of disease should be used as markers for treatment feasibility and outcome.

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Diffuse large B-cell lymphoma (DLBCL) is the most common lymphoid malignancy in adults.

The median age of presentation of DLBCL falls between the sixth and seventh decade and accounts for approximately 31% of all non-Hodgkin lymphoma (NHL) in Western countries and 37% worldwide. Disseminated extranodal disease is less frequent, and one-third of patients have systemic symptoms. Overall, DLBCLs are aggressive but potentially curable malignancies. Cure rate is particularly high in patients with limited disease with a 5-year progression-free survival (PFS) ranging from 80% to 85%. Patients with advanced disease or symptomatic disease have a 5-year PFS around 50%.

Secondary cutaneous involvement is uncommon in DLBCL, although it has been observed in up to 20% of cases in some series. The presence of extensive cutaneous lesions is more often observed in secondary cutaneous DLBCL compared with primary DLBCL, leg type. Although skin lesion characteristics do not differ significantly between primary and secondary, extensive cutaneous lesions are more often observed in secondary cutaneous DLBCL compared with DLBCL, leg type. The presence of multiple skin lesions and time of evolution at presentation were associated with poorer prognosis in secondary cutaneous DLBCL.[1–3]

This case presenting with multiple nodular lesions is relevant for the extensive involvement of soft-tissue and individual nodule size that is not a characteristic seen as often as solitary erythematous nodule or erythematous-violaceous plaques.[4 5]

In the case, we chose to report despite the feasibility of treatment and this disease carrying a good prognosis with adequate chemotherapy, the performance status and large burden of disease of the patient have not allowed her to undergo adequate treatment for her disease. A palliative approach was adopted given the overall patient's condition. Overwhelming disease progression followed leading to her death.

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