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CASE REPORT

Diffuse large B-cell lymphoma: an extensive, ulcerated, fungating cutaneous manifestation

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Introduction

Primary cutaneous large B-cell lymphoma leg type is a rare, distinct disease entity with an aggressive course and a poor prognosis. This article presents the case of a 78-year-old woman who was referred from primary care with a rapidly growing, ulcerated, fungating leg lesion, with no systemic symptoms or nodal disease. Biopsy of the lesion revealed histological features consistent with primary cutaneous large B-cell lymphoma, which was partially responsive to chemotherapy. This case highlights the aggressive presentation of primary cutaneous large B-cell lymphoma. It reiterates the importance of investigating skin lesions in older patients and highlights clinicians' needs to consider non-dermatological differential diagnoses for illness with cutaneous manifestations, especially in light of the COVID-19 pandemic where patients may harbour delayed presentations of serious systemic pathology.

Case report

A 78-year-old woman was referred to the surgical team by her GP with a 6-week history of a rapidly enlarging lesion on her right thigh. The lesion started as a small red nodule. Despite the size of the lesion on admission (Figure 1), the patient was mobile, independent and denied any lower limb neurological deficit. There was unintentional weight loss but no fever or night sweats. The patient denied a history of recent bites or foreign travel. Past medical history included endometrial cancer (pT3a Nx M0) for which the patient had undergone a total abdominal hysterectomy and bilateral salpingo-oophorectomy followed by postoperative pelvic radiotherapy when she was 68 years old. The patient also had hypertension and atrial fibrillation. She was a lifelong non-smoker.

On examination, she appeared well with no peripheral signs of anaemia. Examination of the respiratory, cardiovascular and abdominal systems was unremarkable. On the anteromedial aspect of the right thigh, extending to just above the level of the knee joint, was a large 30 cm long and 15 cm wide fungating lesion with distinct but thick everted edges and an ulcerated centre (Figure 1). The lesion was friable with pinpoint bleeding noted centrally. There was minimal cellulitis surrounding the lesion. Ipsilateral inguinal lymphadenopathy was noted, but no other clinically palpable lymphadenopathy.

Initial observations were all within normal limits. The full blood count demonstrated a normocytic anaemia (haemoglobin 86 g/litre) with white cell count within normal limits. The C-reactive protein level was elevated at 157 mg/litre, and serum lactate dehydrogenase level was 1723 U/litre. HIV test was negative. A staging computed tomography demonstrated enlarged lymph nodes throughout the mediastinum and within the right groin. Lower limb magnetic resonance imaging confirmed a large lobulated mass in the anteromedial aspect of the right thigh measuring 14×10×30 cm (anteroposterior × transverse × craniocaudal) and infiltrating into the overlying skin as well as the underlying musculature (Figure 2).

An excision biopsy was performed. Histological analysis reported a high-grade ulcerating tumour comprising sheets of tumour cells with little cytoplasm and vesicular nuclei with prominent nucleoli. The tumour had a starry sky appearance, with tumour cells stained strongly positive for leucocyte common antigen, CD20, bcl-2, bcl-6 and MUM1. Nearly 100% of the cells were positive for Ki-67.

Following discussion at the haematology cancer multidisciplinary team meeting, the patient was started on the chemotherapy regimen R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone) for diffuse large B-cell lymphoma leg type. The leg lesion regressed significantly upon starting treatment, but 7 months after diagnosis, the disease progressed and the patient eventually died from it.

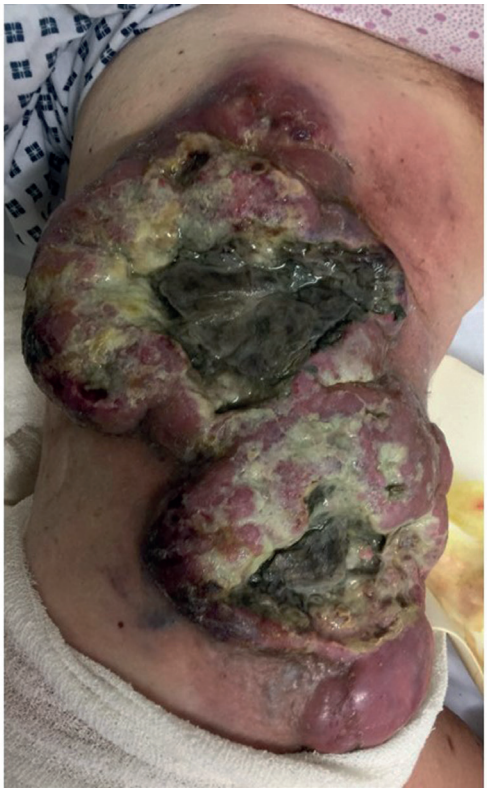

Figure 1. The anteromedial right leg lesion at the time of hospital admission. The lesion extends from the proximal mid-thigh to the level of the femoral condyles.

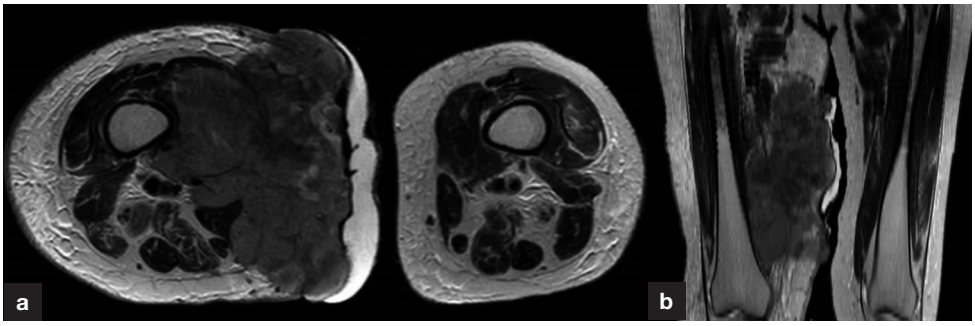

Figure 2. a. Axial and (b) coronal T2-weighted magnetic resonance images of the thigh demonstrating the extensive lesion and involvement of subcutaneous tissues and anterior thigh musculature but absence of bony infiltration.

Discussion

Around 20–30% of primary cutaneous lymphomas are primary cutaneous B-cell lymphomas (Lima, 2015). The pathogenesis of primary cutaneous large B-cell lymphoma leg type has been postulated to be antigenic stimuli in the cutis provoking a lymphoproliferative response (Thomas et al, 2011). Primary cutaneous large B-cell lymphoma leg type is a rare, distinct disease entity (Willemze et al, 2019) that is more common in older women and on the legs (Lima, 2015). The explosive development and aggressive clinical behaviour exhibited in this patient's case are typical of primary cutaneous large B-cell lymphoma leg type (Lipowicz et al, 2011). The absence of other extensive cutaneous lesions makes secondary cutaneous diffuse large B-cell lymphoma unlikely.

Histopathological presence of confluent sheets of large cells with round nuclei defines primary cutaneous large B-cell lymphoma leg type (Grange et al, 2007). The majority of these tumours express Bcl-2. Elevated serum levels of lactate dehydrogenase, MUM-1 expression and positive B-cell markers (CD20 and Ki-67) are other diagnostic features (Willemze et al, 2019).

Learning points

- Primary cutaneous large B-cell lymphoma is a rare non-Hodgkin lymphoma which is more common in older women and on the legs.
- The differential diagnosis for this patient's lesion include a secondary cutaneous involvement, but the isolated nature of this lesion favours primary cutaneous large B-cell lymphoma leg type.
- The overall prognosis of primary cutaneous large B-cell lymphoma leg type is poor, with location on the leg and multiple skin lesions both independent factors associated with a poorer prognosis.
- At present there is no optimal treatment for primary cutaneous large B-cell lymphoma leg type because of the paucity of randomised controlled trials, but the combination of rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone (R-CHOP) remains the recommended treatment regimen.
- The COVID-19 pandemic may cause patients to delay seeking medical advice for skin lesions. Therefore, a thorough systemic assessment and multidisciplinary approach should be adopted by all clinicians encountering patients with such presenting complaints.

The overall prognosis of primary cutaneous large B-cell lymphoma leg type is poor. Leg disease and multiple skin lesions are independent factors associated with a poorer prognosis (Grange et al, 2007). There is no optimal treatment for primary cutaneous large B-cell lymphoma leg type, but R-CHOP remains the recommended regimen (Thomas et al, 2011).

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