Primary cutaneous diffuse large B cell lymphoma leg type mimicking pretibial myxedema

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Introduction

Primary cutaneous diffuse large B cell lymphoma leg type (DLBCLLT) which is a rare subtype of primary cutaneous lymphoma (PCL) accounts for less than 5% of all PCLs. Elderly women are the most commonly affected group[1]. It has an unfavorable prognosis and requires aggressive chemotherapy. Clinical presentation of DLBCLLT encompasses a wide range of possibilities. Here we report a case of DLBCLLT mimicking pretibial myxedema.

Case report

A 70-year-old female was referred to the dermatology clinic, with progressively growing painful skin lesions over bilateral shins for 3 months. She had a history of type 2 diabetes mellitus which was well under control and had been treated for pulmonary tuberculosis at the age of 33. Her history did not reveal any constitutional symptoms and she was clinically euthyroid.

On examination there were ill defined tender erythematous plaques and nodules which were firm in consistency over anterior aspects of both shins. There was no ulceration or discharge. Overlying skin was thickened and hardened. Hair follicles were prominent with *peaud'orange* appearance (Figure 1). There was a diffusely enlarged non tender thyroid goiter. There was no lymphadenopathy or hepatosplenomegaly. Rest of the physical examination was normal.

We considered pretibial myxedema, cutaneous lymphoma, erythema induratum as differential diagnoses.

Full blood count, blood picture, erythrocyte sedimentation rate, lactate dehydrogenase level, T4/TSH and chest radiography were normal. Ultrasonograpgy of the neck revealed a multinodular goiter and of abdomen it was normal.

An incisional skin biopsy revealed an atrophic epidermis, diffuse infiltrate of medium to large lymphoid tissue in dermis with a grenz zone, focally involving the subcutaneous tissue. Cells appear to dissect the collagen. Large cells contain hyperchromatic irregular nuclei with scanty cytoplasm. Mitoses are seen with interspersed small round lymphoid cells (Figure 2).

Immunohistochemistry of large tumor cells showed strong membrane positivity for CD20 (Figure 3) and cytoplasmic positivity for BCL2 (Figure 4). Only a few cells showed MUM1 nuclear positivity. Small cells showed membrane positivity for CD3 (Figure 5). Ki 67 proliferation index was 75%.

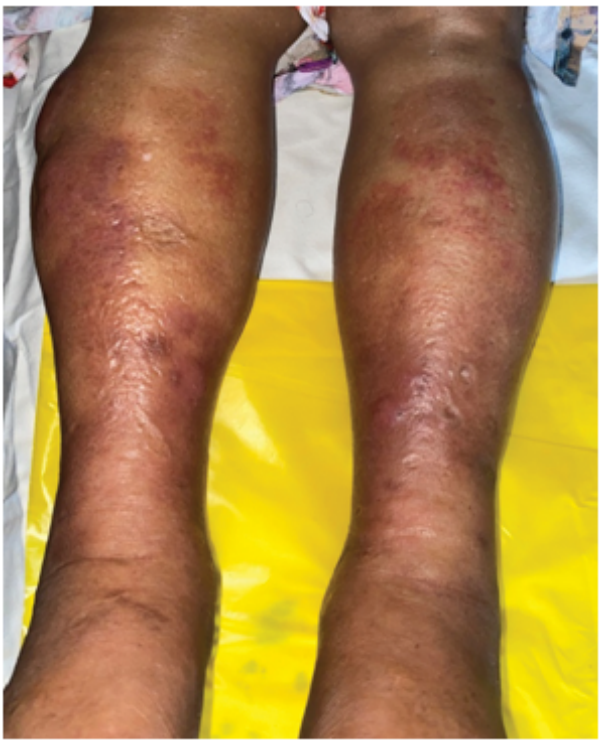

Figure 1.

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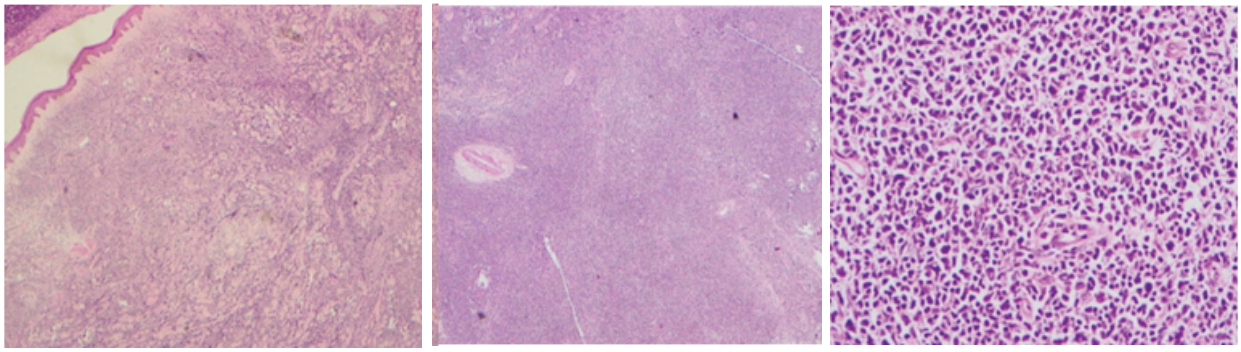

Figure 2.

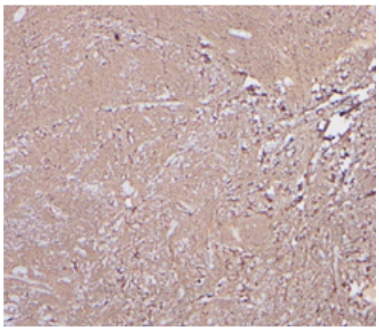

Figure 3.

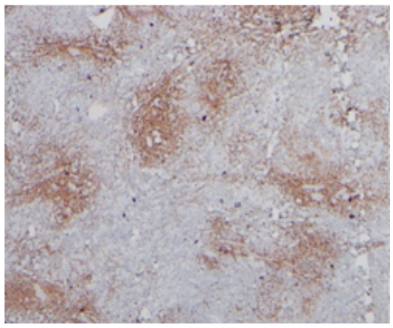

Figure 4.

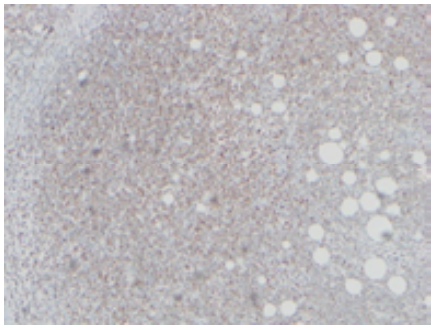

Figure 5.

Subsequent staging investigations including a bone marrow biopsy and a contrast enhanced CT chest, abdomen and pelvis were negative. A diagnosis of primary cutaneous diffuse large B cell lymphoma, leg type was made and patient was referred to haemato-oncology team for the further management.

Discussion

Primary cutaneous diffuse large B cell lymphoma is an extra nodal non hodgkin lymphoma which accounts for 4% of all cutaneous lymphomas and 20% of all primary cutaneous B cell lymphomas. According to WHO EORTC classification there are three subtypes of PCBCL (Primary cutaneous B cell lymphoma (PCBCL) has three main subgroups. Primary cutaneous marginal zone lymphoma (PCMZL), primary cutaneous follicle center lymphoma (PCFCL) and primary cutaneous diffuse large B-cell lymphoma, leg type (DLBCLLT)[2,3]. Among them DLBCLLT is the rarest subtype with increased rates of relapses and poor prognosis. It preferentially affects elderly population with average age of 78 and has a female preponderance[1]. It has various clinical presentations making the diagnosis challenging.

It presents as solitary or multiple rapidly growing nodules and plaques which frequently ulcerate[1]. Legs are involved in 80% of cases. It can involve both legs and resembles the clinical picture of pretibial myxedema like in our patient. 40% of cases have extracutaneous progression with bone marrow and lymph node involvement[5]. About 50% of cases have a recurrence. 5 year survival rate reported to be only 58%.

Clinicopathological and immunohistochemical analysis together aid in arriving at the diagnosis of DLBCLLT.

Histology shows dense infiltrate of monomorphic B cells with large cell morphology extending to deep dermis and subcutaneous tissue[4]. There are no reactive or inflammatory cells in the background. B cell markers such as CD12, CD20, CD22, CD79a are expressed in the neoplastic cells. Expression of Bcl-2, Bcl-1, MUM1 and FOXP1 markers are characteristic of DLBCLLT.

Recommended treatment options include aggressive systemic therapy with combination chemotherapy with or without radiotherapy. Further rituximab is considered as an alternative therapy for elderly with poor tolerance to chemotherapy[6].

Conclusion

Primary cutaneous lymphoma is a diagnostic challenge which has various clinical presentations.

Our case highlights the importance of knowing the classic as well as unusual presentations of DLBCLLT. In the presence of clinical characteristics such as advanced age, lower limb involvement, rapidly evolving skin lesions DLBCLLT should be considered as a differential diagnosis.

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