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Case Report

Subcutaneous panniculitis-like T cell lymphoma: a rare entity and its cryptic journey through pregnancy

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ABSTRACT

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare form of skin lymphoma that primarily is localized to the subcutaneous adipose tissue and accounts for less than 1% of all peripheral T-cell lymphomas. It presents with multiple subcutaneous nodules or plaques on extremities and has poor prognosis if accompanied by hemophagocytic syndrome. Differential diagnosis is panniculitis, lupus panniculitis and leprosy. We report such a rare case of a female with lupus erythematosus (LE) panniculitis like presentation with favourable outcome to oral steroids in pregnancy.

Keywords: Subcutaneous panniculitis like T cell lymphoma, Pregnancy, Lupus like presentation

INTRODUCTION

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare form of cutaneous T-cell lymphoma characterized by neoplastic αβ T cells infiltrating subcutaneous tissues in a lobular pattern.[1] Patients typically have indurated and sometimes erythematous plaques and nodules, often on the lower extremities, although the upper extremities and the face can be involved. There is no standardized treatment protocol for SPTCL, and even fewer data guide treatment for patients diagnosed during pregnancy. SPTCL is rare but its vital role for dermatologists to be aware of this entity due to its clinical and histological resemblance to lupus panniculitis.[2]

CASE REPORT

A 35-year-old female presented with chronic recurrent febrile episodes with chills since past 7 years associated with multiple swellings over the lower extremities which gradually progressed to involve, upper extremities, abdomen and face. Fever and swellings would subside partially on taking antipyretics and systemic steroids and lesions would reappear when medication was being tapered off. Patient was diagnosed as Hansen's disease with type 1 reaction in the past and had received MB-MDT for 1 year with no improvement in the lesions. Patient also gives history of pregnancy, detected during 5th month of gestation, with favourable outcome with oral steroids. She also gives history of photosensitivity, recurrent painful oral ulcers, shortness of breath, proximal muscle weakness and hair loss. On examination patient appear pale, had bilateral pitting pedal oedema extending till ankle, bilateral non tender inguinal lymphadenopathy and hepatosplenomegaly.

On cutaneous examination the patient had generalized involvement in the form of indurated plaques and nodules typically over the face, upper extremities, lower extremities and abdomen and breast. Few lesions were tender and ranging in size from 1 cm to 8 cm in diameter (Figure 1 and 2). There was no peripheral neuropathy. There was decreased density of hair over the scalp and hair pull test was negative.

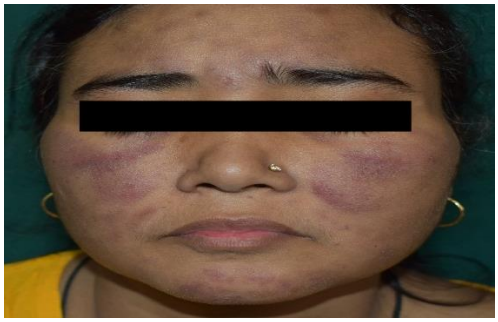

Figure 1: Erythematous indurated plaques over face.

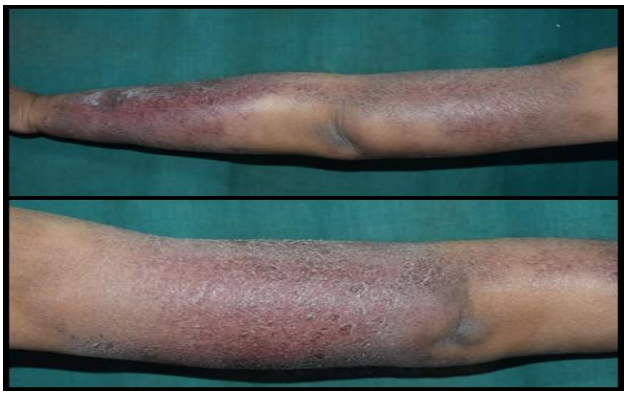

Figure 2: Multiple erythematous indurated plaques over extensor aspect of bilateral upper extremities.

Routine laboratory investigations revealed anaemia (haemoglobin 9.8 g/dl), leukopenia and deranged liver function tests. Renal function tests were normal. Chest X-ray was normal.

Based on the above differential diagnosis of systemic lupus erythematosus, panniculitis and lupus panniculitis were kept.

On further workup, antinuclear antibody titres were normal and antibodies to double stranded deoxyribonucleic acid (DNA) were absent, low complement levels, high creatine phosphokinase-MB (CPKMB), high lactate dehydrogenase (LDH), anti-smooth muscle antibody weakly positive and high angiotensin converting enzyme (ACE) inhibitor. Ultrasonography abdomen, pelvis and breast showed hepatosplenomegaly, fatty liver change and fat necrosis in breast respectively. Histopathological examination of lesions showed lymphomatous infiltrate involving predominantly the subcutaneous lobules in a lobular panniculitis like pattern. Dermis and epidermis were spared (Figure 3a). On higher magnification, the lymphoid infiltrate was diffuse and had enlarged and hyperchromatic nuclei, occasional mitosis, karyorrhexis and rimming of adipocytes by these atypical neoplastic lymphoid cells (Figure 3b).

Immunohistochemical evaluation showed CD3 and CD8 positivity while CD30 and CD56 negativity (Figure 4a and b). Bone marrow examination was normal. Positron emission tomography (PET) scan showed metabolically active diffuse cutaneous thickening with associated subcutaneous fat stranding associated with bilateral inguinal, external iliac and femoral adenopathy.

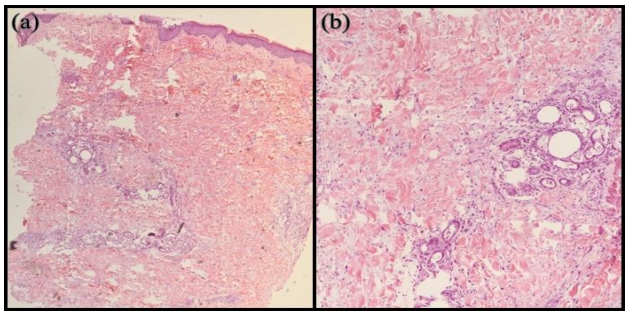

Figure 3: (a) Hematoxylin and eosin stain on 4x magnification showing lymphomatous infiltrate involving predominantly the subcutaneous lobules in a lobular panniculitis like pattern with sparing of epidermis and dermis; (b) on 10X magnification, the lymphoid infiltrate was diffuse and had enlarged and hyperchromatic nuclei, occasional mitosis, karyorrhexis and rimming of adipocytes by these atypical neoplastic lymphoid cells.

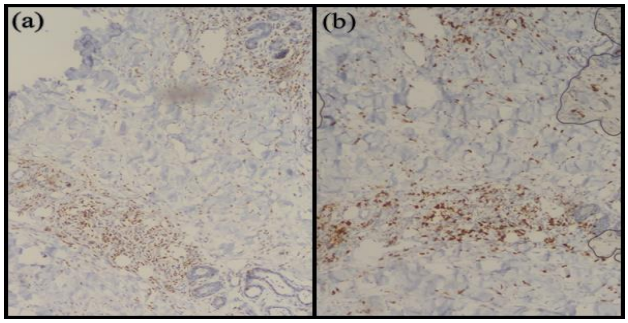

Figure 4: Immunohistochemical stain showing (a) CD3 positivity and (b) CD8 positivity.

Based on above final diagnosis of subcutaneous panniculitis like T cell lymphoma was made.

The patient was referred to haematology department and she underwent five cycles of CHOP chemotherapy regimen (cyclophosphamide 1220 mg, doxorubicin 81 mg and vincristine 2 mg along with oral prednisolone). Patient is in remission since then.

DISCUSSION

SPTCL is a rare distinct primary cutaneous lymphoma characterized by infiltration of subcutaneous tissue by neoplastic cytotoxic alpha and beta T cells mimicking panniculitis.[1] Gonzalez et al was the first to describe it in 1991 when they presented with 8 cases of T cell lymphoma with subcutaneous involvement. In 2001, World Health Organization (WHO) classification included it as a distinct entity. Incidence is less than 1% of non-Hodgkin's lymphomas. It affects both children

and young adult with mean age of onset being 36 years and showing female preponderance. 20% of cases are associated with an autoimmune aetiology especially lupus erythematosus. According to some authors, both the entities lie on a same disease spectrum and when overlapping features are present it is termed as atypical lymphocytic lobular panniculitis. It is hypothesized that migration of neoplastic T cells to adipocytes is facilitated by CCR5 expression.[2]

SPTCL presents with multiple painless erythematous subcutaneous plaques and nodules typically on lower extremities, upper extremities and trunk. Lesions have a waxing and waning course.[3] Systemic symptoms like fever, chills, myalgias, cytopenia and deranged liver function test are seen in 50% of cases, more common in patients with concurrent hemophagocytic syndrome. Lymph node and bone marrow involvement is very rare.[4]

Histopathology of skin lesions reveal dense lymphoid infiltrate of small to large size lymphocytes in the subcutaneous tissue in lobular pattern. The lymphocytes are atypical with hyperchromatic angulated nuclei and irregular borders, occasional mitoses and karyorrhexis. Interlobular septa, dermis and epidermis are usually spared. These lymphocytes are usually seen rimming adipocytes with fat cell necrosis which gives a diagnostic clue. On immunohistochemistry, these cells show CD8, perforin, granzyme B and TIA1 positivity.[5]

There is no standardized treatment protocol for SPTCL. Most of the cases respond successfully to systemic corticosteroids or immunosuppressants like cyclosporine A, methotrexate, chlorambucil and bexarotene. In progressive disease, there is a role of conventional doxorubicin-based chemotherapy. Chemotherapy with cyclophosphamide, doxorubicin, vincristine, and prednisolone is the preferred regimen, with overall remission rates of 50%.[6] There have been reports about remission of SPTCL with oral corticosteroids during pregnancy as seen in our case.[7] Radiotherapy has palliative role and may induce long term remissions. In refractory cases, stem cell transplant is the last resort.[8] Many cases have excellent prognosis with 5-year survival rate of 85-91%. HPS and upper extremity involvement have worse prognosis.[9]

There are only few case reports of SPTCL in pregnancy and there is no standard treatment protocol of SPTCL. Long term follow-up and further studies are required to know the course of the disease.

CONCLUSION

SPTCL is rare cutaneous lymphoma, and exceptionally rare during pregnancy. It has a tendency to be misdiagnosed as LE panniculitis due to their clinical and histological similarities, which eventually leads to delay in the diagnosis and further progression of disease. It is important to diagnose this condition early and confirming the same with immunohistochemistry in case of strong suspicion. Stabilisation of the disease with oral corticosteroids during pregnancy is the key. Multidisciplinary approach is needed for managing patients with this condition.

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