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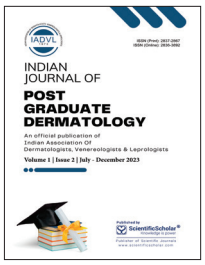

Clinicopathologic Challenge

Intriguing Subcutaneous Nodules – A Panic-Stricken Panniculitis

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ABSTRACT

A 13-year-old boy presented with multiple transient painless swellings over the body for the past 6 months. Lesions would persist for 6–8 weeks and spontaneously resolve leaving behind discolouration and altered skin texture. Clinical examination revealed a single soft, non-tender mobile nodule of 2 × 2 cm over left arm, along with multiple discrete well defined post-inflammatory hypopigmented patches with hyperpigmented scaly papules over the arms, thighs and trunk. Skin biopsy showed lobular panniculitis composed of lymphoid cells, histiocytes and plasma cells with characteristic rimming of fat cells by lymphoid cells suggestive of cytophagic histiocytic panniculitis. Immunohistochemistry showed positive CD3, CD5, CD7 and CD8, negative CD20, CD4, CD56, Ki67 – 75–80%, CD68 highlighting the histiocytes and C30 negative. Positron emission tomography–magnetic resonance imaging showed multiple fluorodeoxyglucose (FDG) avid cutaneous and subcutaneous nodules throughout the body and minimally FDG avid axillary and inguinal lymph nodes. Thereby, a final diagnosis of subcutaneous panniculitis like T-cell lymphoma was concluded. Our patient was started on a tapering dose of oral prednisolone and cyclosporine, following which the lesions significantly improved.

Keywords: Subcutaneous, Nodule, Panniculitis, T-cell lymphoma

INTRODUCTION

Panniculitis refers to the inflammation of the subcutaneous fat. It often poses diagnostic difficulties, both clinically as well as histopathologically. Here, we report a child with intriguing subcutaneous nodules, who on further evaluation, was found to have a rare cutaneous T-cell lymphoma.

CASE REPORT

A 13-year-old boy with no known comorbidities presented with multiple transient painless swellings over the body for past 6 months. Lesions would persist for 6–8 weeks and resolve spontaneously leaving behind discolouration and altered skin texture. There was no history of associated fever or joint pains. Clinical examination revealed a single soft and non-tender mobile nodule of 2 × 2 cm over the left arm along with multiple discrete well-defined post-inflammatory hypopigmented patches with hyperpigmented scaly papules over the arms, thighs and trunk [Figure 1]. Diffuse oedema over the bilateral arms and forearms present. No palpable lymph nodes or sensory loss noted. There were no features of systemic involvement. Clinical differential diagnoses included panniculitis (various causes including erythema nodosum, infections, connective tissues disease, sarcoidosis and pancreatic panniculitis), vasculitis, lymphoproliferative disease and Hansen's disease.

Routine blood investigations showed leukopenia, mildly elevated inflammatory markers, borderline transaminitis and mildly elevated lipase level. Immunofluorescence for anti-nuclear antibody (ANA) showed borderline fine speckled pattern; however, ANA profile was negative. An ultrasonography abdomen showed hyperechoic lesions in the right flank and epigastric region in subcutaneous plane.

Skin biopsy showed lobular panniculitis composed of lymphoid cells, histiocytes and plasma cells with characteristic rimming of fat cells by lymphoid cells suggestive of cytophagic histiocytic panniculitis [Figure 2a-c]. Fat necrosis, fibrinoid deposition and apoptotic debris were also seen. The differential diagnoses were narrowed down to reactive haemophagocytosis, subcutaneous panniculitis like T-cell lymphoma (SPTCL) and lupus erythematosus panniculitis. Immunohistochemistry (IHC) showed positive CD3, CD5, CD7 and CD8, CD68 highlighting the histiocytes, Ki67 index 75–80% [Figure 3] and negative CD20, CD4, CD56 and C30.

Positron emission tomography-magnetic resonance imaging showed multiple fluorodeoxyglucose (FDG) avid cutaneous and subcutaneous nodules throughout the body [Figure 4] and minimally FDG avid axillary and inguinal lymph nodes. Lymph node biopsy showed lymphohistiocytic infiltrate in perinodal fat with CD8 positive lymphocytes and CD68 positive histiocytes in IHC. Bone marrow study showed variably cellular marrow showing evidence of haemophagocytosis.

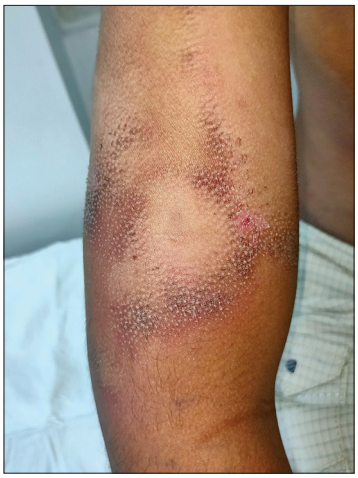

Figure 1: Subcutaneous nodule over left arm with post inflammatory hypopigmented patches and peripheral hyperpigmented scaly papules.

Thereby, a final diagnosis of SPTCL was concluded. Our patient was started on a tapering dose of oral prednisolone and cyclosporine, following which the lesions significantly improved. He is on regular follow-up and monitoring.

DISCUSSION

Originally described by Gonzalez *et al*. in 1991, SPTCL is a rare T-cell lymphoma of the skin contributing to <1% of all non-Hodgkin's lymphomas.[1]

The new World Health Organisation (WHO)-European Organisation for Research and Treatment of Cancer (EORTC) classification for cutaneous lymphomas considers SPTCL a separate entity. Based on the T-cell receptor (TCR) and immunophenotype, it is classified into two types: (a) α/β: Indolent course with CD4–, CD8+ and CD56– and 5-year survival rate of 82% and (b) γ/δ: Epidermal involvement with an aggressive and fatal course due to associated haemophagocytosis syndrome with CD4–, CD8– and CD56+ and 5-year survival rate of 11%. In the WHO-EORTC classification, SPTCL now refers to cases with an α/β TCR phenotype, whereas a new category named cutaneous γ/δ T-cell lymphoma includes cases with a γ/δ TCR phenotype.[2]

The usual clinical presentation of asymptomatic subcutaneous nodules mimics other benign causes of panniculitis, and hence, diagnosis may be missed if histopathological examination is not thoroughly evaluated. Lobular panniculitis with a variably dense infiltrate of atypical CD8+ T-lymphocytes rimming the adipocytes is characteristic of SPTCL. The neoplastic T-lymphocytes next to the adipocytes have a higher Ki67 index. Fat necrosis, karyorrhexis and mitoses are commonly seen.[3]

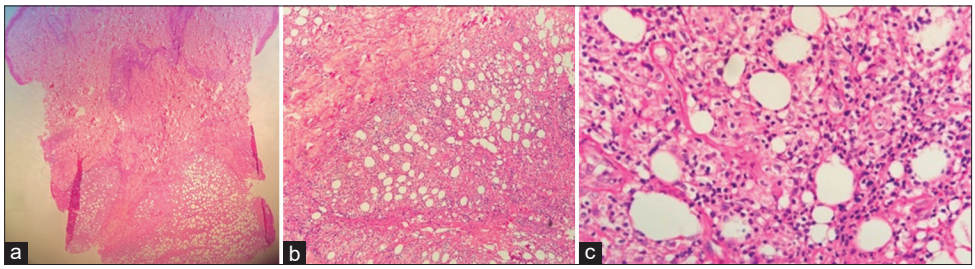

Figure 2: (a) Scanner view showing panniculitis (Haematoxylin and Eosin, ×40). (b) Lobular panniculitis with lymphoid cells, histiocytes and plasma cells (Haematoxylin and Eosin, ×100). (c) Characteristic rimming of fat cells by lymphoid cells (Haematoxylin and Eosin, ×400).

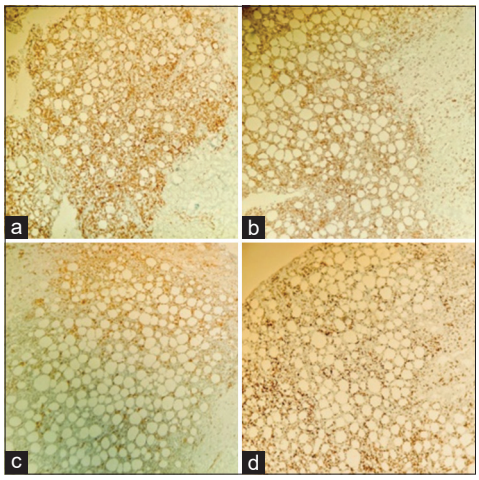

Figure 3: Immunohistochemistry showing positive (a) CD3 staining (×100) (b) CD5 staining (×100) (c) CD68 staining (×100) (d) Ki67 – 75–80% (×100).

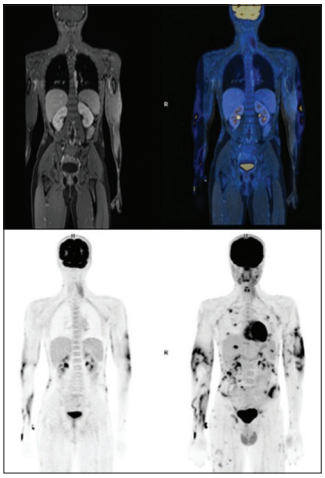

Figure 4: Positron emission tomography-magnetic resonance imaging showing multiple fluorodeoxyglucose avid cutaneous and subcutaneous nodules throughout the body.

Whereas, lupus panniculitis shows interface changes, CD4 and CD8 T-cells without atypia, B-cell and plasma cell aggregates, plasmacytoid dendritic cells and intradermal mucin with a low Ki67 index.[3] Other differential diagnoses like NK/T-cell lymphomas show a neoplastic infiltrate extending into the dermis and epidermis along with angioinvasion, Epstein-Barr virus sequences and CD56 positivity, while anaplastic large cell lymphomas depict epidermal ulceration and CD30 positivity.[4]

SPTCL is best managed by an interprofessional team. Treatment may include systemic steroids, multidrug chemotherapy or cyclosporine. Our patient showed a good response to tapering oral steroids and cyclosporine.

CONCLUSION

Panniculitis remains a challenge for diagnosis. Due to overlapping features with non-neoplastic panniculitides as well as other cutaneous T-cell lymphomas, SPTCL proves to be a diagnostic dilemma.[5] A combination of clinicopathology and IHC is key in the diagnosis of this rare entity which would aid in appropriate treatment and prevent complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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