Nodular enigma: A rare cause of skin nodules in an adolescent boy

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Introduction

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) occurs mainly in adults and is very rare in children[1]. Panniculitis may pose challenges in both clinical and histological diagnosis, due to multiple aetiologies and homogeneous morphological features[1]. We report a 15-year-old boy who presented with raised painful lesions in the skin, and our arduous path to arrive at the diagnosis.

Case report

A 15-year-old boy, born to non-consanguineous parents, presented to us with a 4-month history of fever, nodular skin lesions and weight loss. The skin lesions started as a single, raised, painful lesion in the left thigh which progressed to multiple lesions over the trunk and lower limbs. Some of the lesions were reported to have healed spontaneously over a period of 2 weeks leaving an indurated hyperpigmented scar. The maximum temperature was 104^0F with spikes occurring 6-8 hourly and he lost 12% of his initial weight over a period of 4 months. There was no significant hospitalisation or illness in the past. Child had an uneventful perinatal period and growth and developmental milestones were appropriate as per mother. There was no significant family history.

Clinical examination revealed pallor and pedal oedema; there was no lymphadenopathy or external anomalies. His weight was 32.5kg (<-3Z score) and height was 158cm (between 0 and +2 Z score). He had generalized tender subcutaneous nodules sparing the face, palms and soles. The nodules were in different stages of evolution with concurrent active, healing and healed lesions (Figures 1-3).

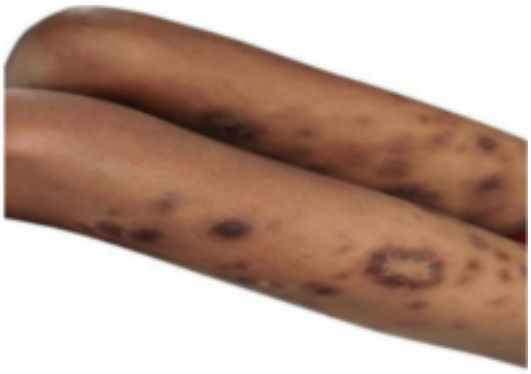

Figure 1: *Multiple firm cutaneous nodules with varying erythema, hyperpigmentation and desquamation of overlying skin in both thighs*

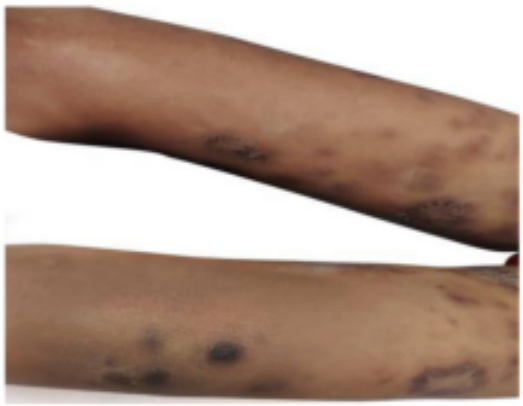

Figure 2: *Nodular lesions along with annular hyperpigmented scales in waxing and waning pattern in both thighs*

Figure 3: *Nodular skin lesions in waxing and waning pattern in upper limb*

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Fine needle aspiration cytology of nodules was reported as consistent with features of erythema nodosum (EN) elsewhere. Empirical treatment with steroids for 2 weeks and antitubercular therapy for 4 weeks were given without significant improvement. Chest, cardiovascular examination and neurological examination were unremarkable; there was no hepatosplenomegaly on clinical examination.

Further investigations showed pancytopenia [haemoglobin 8.6g/dl, white blood cell count 2.4 x 10^9/l (polymorphs 34%, lymphocytes 59%, eosinophils 4%, monocytes 2% and basophils 1%) and platelet count 139 x 10^9/l]. His erythrocyte sedimentation rate was 12mm/hour and C-reactive protein level was 5.6mg/L (normal <10mg/L). The serum ferritin level was 950ng/ml (normal:15-135ng/ml) and the serum albumin was 2.5 g/L (normal <3g/L). Bone marrow study, done in view of the pancytopenia, showed preserved trilineage haematopoiesis with haemophagocytosis. Antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA) were negative.

A repeat biopsy from skin nodules revealed diffuse, predominantly lobular infiltrates of atypical lymphoid cells in the subcutaneous fibroadipose tissues rimming the fat spaces (Figure 3: A to D).

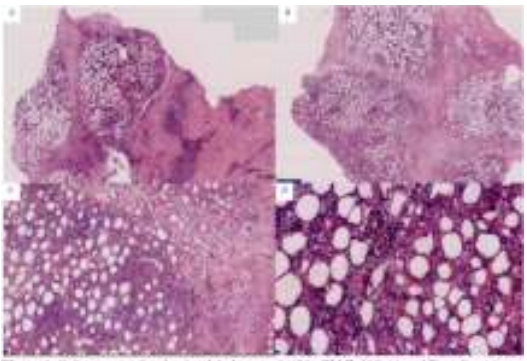

Figure 4: *Subcutaneous fat showing lobular panniculitis (H&Ex100) (A and B), subcutaneous fat showing rimming of the fat globules by lymphoid cells (H&Ex200) (C) and (H&Ex400) (D)*

Immunohistochemistry (IHC) was positive for CD3 /CD8 /CD68 /CD2 /CD7 /Granzyme-B. Furthermore, polymerase chain-based assay was done on clonal re-arrangement, which was negative for gamma chain re-arrangements. A diagnosis of subcutaneous panniculitis-like T-cell lymphoma (SPTCL) was made. The patient was started on CHOP chemotherapy regimen (cyclophosphamide, doxorubicin, vincristine and prednisolone). Marked clinical resolution in skin nodules and fever was observed after the start of chemotherapy.

Discussion

Panniculitis encompasses varying groups of inflammatory diseases that involve subcutaneous adipose tissue with similar clinical presentation regardless of the aetiology with erythematous, mostly tender subcutaneous nodules in areas of prominent fatty tissue like the thighs, buttocks and legs[2]. EN is classically a septal panniculitis. Predominant lobular panniculitis in our patient was unlikely for EN[2].

SPTCL is a rare primary cutaneous lymphoma and accounts for less than 1% of non-Hodgkin lymphomas. It is associated with lupus erythematosus in 20% cases and is an important differential diagnosis due to its similar clinical and histological course[3]. SPTCL is a lobular panniculitis with dense nodular infiltration of pleomorphic lymphocytes in the subcutaneous fat. The cells are typically seen rimming individual adipocytes, as seen in our patient. Aggregates of atypical lymphocytes, along with necrotic lymphocytes, also known as ghost cells, and apoptosis with karyorrhectic debris are often linked with the presence of haemophagocytic cells. Paediatric lesions are frequently associated with plasma cells. Methodical examination of multiple serial sections of the skin biopsy is needed for a definitive diagnosis[4]. Immunohistochemistry (IHC) indicates that the lymphocytes are mostly cytotoxic CD8+ cells expressing cytotoxic proteins granzyme B, but not CD56[4]. Typical sparing of epidermis / dermis helps to differentiate it from lupus panniculitis and rimming of individual fat cells by neoplastic T-cells was a helpful diagnostic feature[5]. Haemophagocytosis in SPTCL is seen in 15-20% cases and is associated with a poor prognosis[4].

Being a disease primarily presenting with cutaneous symptoms, patients approach dermatologists first, and it is imperative for them to be aware of the entity along with other differential diagnoses. Empirical use of steroids can mask the diagnosis of rare entities, and needs to be reconsidered without appropriate evidence.

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