Subcutaneous panniculitis like T cell lymphoma

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Sri Lanka Journal of Dermatology, 2006, **10**, 40-41

Abstract

A 22 year old female presented with erythematous tender nodules on trunk and limbs for 6 weeks and associated fever, malaise, and loss of appetite for 2 weeks. She had a high ESR, leukopenia and thrombocytopenia.

Histological examination revealed lobular panniculitis with atypical lymphocytic infiltrate. Cytophagocytic histiocytes were not seen. Immunohistochemical studies confirmed a T cell phenotype.

Subcutaneous panniculitis like T cell lymphoma (SPTCL) was diagnosed. Chemotherapy was commenced and the initial response was satisfactory.

T cell lymphoma, though rare, has to be included in the differential diagnosis of sub cutaneous nodules. Increased awareness of this rare disease would help in early diagnosis of this non specific common presentation.

Introduction

Subcutaneous panniculitis like T cell lymphoma was first described in 1985. It is now included in WHO – EORTC classification of cutaneous T cell lymphoma as a rare subtype. The disease is known to have two clinical presentations[1]. One with an indolent form with good prognosis and the other with rapid clinical detoriation mostly secondary to the development of haemophagocyte syndrome. Haemophagocytic syndrome is characterized by the proliferation of histiocytes and phagocytosis of blood elements, hepatosplenomegaly, pancytopenia and coagulopathy.

Case History

A 22 year old female presented with painful red lesions on lower abdomen, thighs and arms of 6 weeks duration. It was associated with low grade fever, malaise and loss of appetite for the last 2 weeks.

On examination she had a temperature of 101° F and multiple tender erythematous subcutaneous nodules and plaques on lower abdomen, thighs and arms.

She had no lymphadenopathy, mucocutaneous bleeding or hepatosplenomegaly.

Laboratory investigations were as follows. ESR 110mm 1st hour, hemoglobin 12g/dl, leukopenia with a white cell count of 3.2×10^3 and a differential count of 64% of neutrophils, 11% of lymphocytes, 10% of eosinophils. Platelet count was 104,000/mm^3 . Blood picture showed normochromic and normocytic red blood cells, leukopenia and thrombocytopenia. Atypical cells were not seen Her blood culture was negative, liver transaminases were marginally raised. Serum proteins were normal. Renal function and coagulation studies were normal. ANA, HIV and EBV serology were negative.

Histological examination of an excised nodule revealed normal epidermis and dermis. The subcutaneous tissue showed a lobular panniculitis with atypical lymphocytes rimming the adipocytes. There were no cytophagocytic histiocytes in the tissue section.

The immunohistochemical studies revealed the atypical lymphocytes to be of T cell lineage with positive CD_3 and CD_8 markers.

Bone marrow showed no abnormality. The diagnosis of subcutaneous panniculitis of T cell lymphoma was made. The patient was started on chemotherapy.

A remarkable improvement of the clinical picture was seen with disappearance of systemic symptoms and subcutaneous nodules. Currently the patient is undergoing chemotherapy cycles (3 months following the diagnosis).

Discussion

SPTCL is a rare disease. Sometimes it may resemble benign forms of panniculitis both clinically and histopathologically[2].

It presents as crops of subcutaneous nodules in peripheral limbs and trunk Ulceration is uncommon. It may be associated with systemic symptoms as fever, loss of appetite, and loss of weight. Dissemination to extracutaneous sites is rare. Our patient presented with multiple tender subcutaneous nodules. This is a common form in all forms of panniculitis.The rapid progression of the skin lesions, systemic symptoms and abnormal blood count made us think of this rarity.

The careful examination of the histopathological section is important in the diagnosis. Usually the

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epidermis and the dermis are normal. In the early stage focally atypical lymphocytes will be seen and it will mimic other forms of lobular panniculitis. Cases without atypical cells in the early stage of the disease are reported highlighting the need for repeated biopsy and continuous follow up if this disease is suspected[3]. The typical histological picture is a predominantly lobular panniculitis with large atypical lymphocytes with large hyperchromatic nuclei and mitotic figures rimming the adipocytes (Figure 1 and 2). Cytophagocytic histiocytes may also complicate this picture in some.

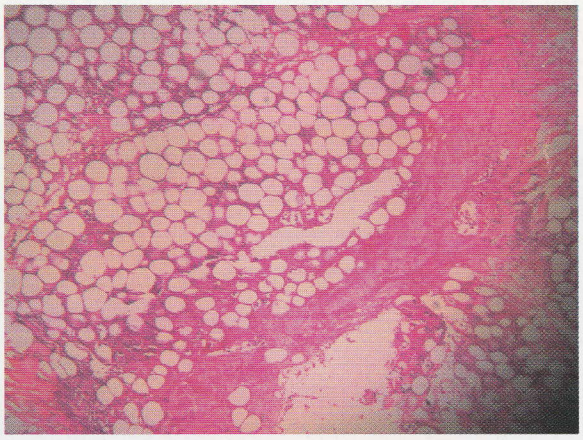

Figure 1.

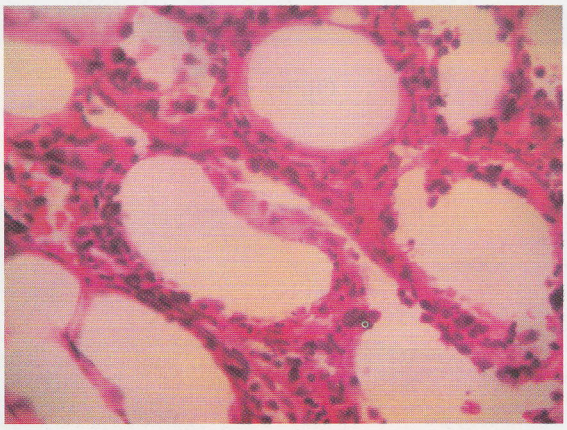

Figure 2.

The immunohistochemical studies will identify these atypical cells as of T cell lineage with positive CD_3 and CD_8. Clonal rearrangement of T cell receptor gene is documented[3].

The prognosis of the disease is poor if they develop the complication of Haemophagocytic syndrome. The patients will be unwell with fever. hepatosplenomegaly, cytopenia and disseminated intravascular coagulation. Fatal haemorrhage and infection will lead to death.

Our patient had fever and bicytopenia suggesting a possibility of a haemophagocytic syndrome. But this was not confirmed with investigations. The skin biopsy did not reveal any cytophagic histiocytes and the bone marrow biopsy was normal.

In the absence of haemophagocytic syndrome SPTCL is treated with chemotherapy (CHOP or CHOP based). Cyclosporine may be the initial treatment when it is associated with haemophagocytosis[4]. Response to treatment is relatively poor specially when associated with haemophagocytic syndrome. But early diagnosis and treatment as in our patient well results in good clinical outcome. High dose chemotherapy is considered with autologus peripheral stem cell transplantation in a relapse[4].

Panniculitis is a relatively common problem in dermatological practice. The reflection of a rare malignant disease in a common presentation is highlighted in this case.

References

1. Charles Perniciaro, Mark J, Jhon White W, et al. Subcutaneous T Cell Lymphoma. *Archives of Dermatology* 1993; **129**: 1171-6.
2. Peter von dev Driesch, Gyde Staib, Miklos Simon. Subcutaneous T cell lymphoma. *Journal of the American Acadamy of Dermatology* 1997; **36**: 285-9.
3. Roger H Weening, Christian S, Charles Pernicaro. *American Journal of Dermatopathology* 2001; **23**: 206-15.
4. Nathaliya N Nguyen. *Dermatology online Journal* **9** (14): 42.