Case Report

Subcutaneous Panniculitis like T cell Lymphoma: A case report.

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Abstract

Sub cutaneous panniculitis like T- cell lymphoma (SPTCL) is a distinct cutaneous T- cell lymphoma characterized by primary involvement of the subcutaneous fat in a manner mimicking panniculitis. We report here a 32 years old male presenting with multiple painful sub cutaneous nodules over extremities and trunk which later healed with atrophic patches. The histopathological finding from the nodules were dense infiltration of atypical lymphoid cells and medium size vessel vasculitis. Immunohistochemistry report showed LCA (CD 45) positive, CD 3 positive, CD4 positive, CD8 positive and CD 20 negative and Cd 56 negative.

Keywords: Panniculitis, T- cell lymphoma, Immunohistochemistry.

Introduction:

Sub cutaneous panniculitis like T- cell lymphoma (SPTCL) was first described by Gonzalez and co-workers in 1991 characterized by an aggressive clinical course and hypodermal involvement [1]. This represents less than 1% of non-Hodgkin's lymphoma which usually affects younger adults with an equal sex incidence and resembles certain benign panniculitis like lupus panniculitis and erythema nodosum clinically and histologically[2]. Histologically there is a lacelike infiltration of the lobules of adipocytes, mimicking panniculitis, a characteristic feature is riming of neoplastic cells around individual adipocytes with nuclear molding and atypia. Malignant cells are predominantly cytotoxic T cells with expression of αβ of T cell receptor (TCR). [3]

Case report:

A 32-year-old male presented with a 16 years' history of multiple painful nodules of varying sizes over both forearms, lower legs and right side of the trunk those healed with central depression with surrounding hyper pigmented border. Initially erythematous painful swelling appeared over the forearm near the wrist joint which increased in size gradually and became hyper pigmented and then healed with depression and residual hyperpigmentation. With time he developed same type of lesions over trunk and lower legs. There was a history of recurrent low grade fever, fatigue but no weight loss or any other systemic symptoms. Examination revealed solitary non tender firm nodule with overlying erythema over the left side of the upper trunk; multiple atrophic plaques of different size and shape with smooth shiny surface and surrounding hyper pigmented well-defined border over both forearms, lower legs and lateral trunk (figure 1a and 1b).

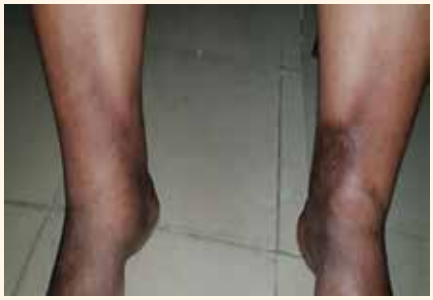

Figure: 1a

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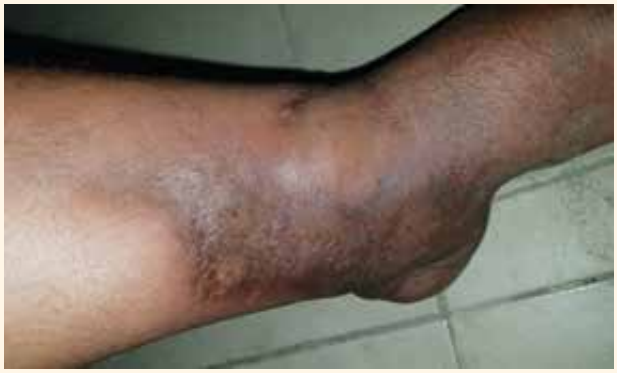

Figure: 1b

Hair, nail, oral and genital mucosa were normal. General examination revealed nothing abnormality with no lymphadenopathy. Systemic examination was normal and no organomegaly was present. Skin biopsy for histopathology was performed and epidermis shows hyperkeratosis, mild acanthosis with mild vacuolar degeneration of the basal layer cells. The subcutaneous tissue shows dense infiltration of atypical lymphoid cells and medium size vessel vasculitis. The upper dermis shows dense perivascular infiltration of chronic inflammatory cells.

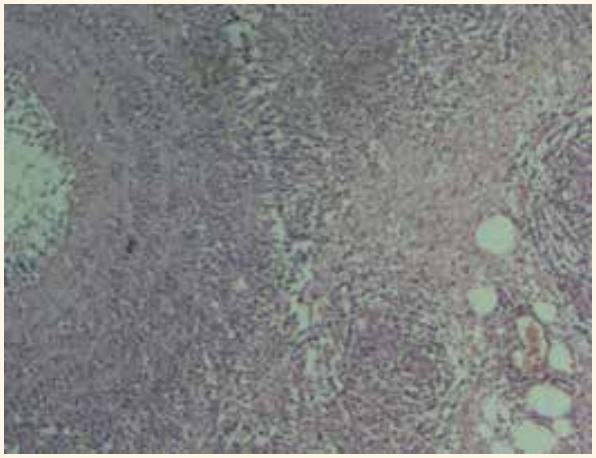

Figure: 2a

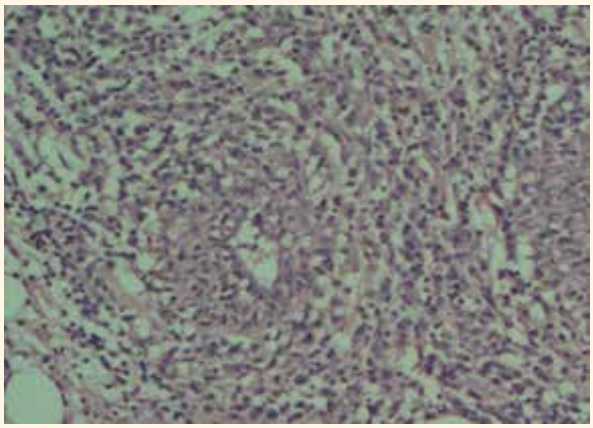

Figure: 2b

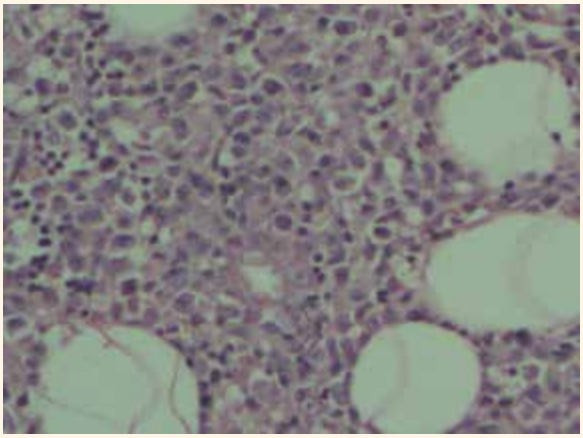

Figure: 2c

Immunohistochemistry showed the cells were CD45+, CD3+, CD20-, CD8+, CD56-, CD4 a few cells positive. The diagnosis of subcutaneous panniculitis like T cell lymphoma done on the basis of clinical features, histopathology and immunophenotypings (Figure-2a, b, c).

Discussion:

Subcutaneous panniculitis like T cell lymphoma is a cytotoxic T-cell lymphoma. It is a rare entity and the WHO-EORTC classification now defines SPTCL as a tumor that expresses α/β T-cell receptor (TCR) gene rearrangement. Tumors expressing γ/δ TCR are classified as primary cutaneous γ/δ T-cell lymphoma. The subcutaneous panniculitis like T cell lymphoma occurs in both children and adults. The median age at diagnosis is 46.5 years, and about 20% of patients are younger than 20 years.[4, 5]The underlying molecular pathogenesis remains yet to be established. Recent studies have found a specific pattern of chromosomal abnormality which have shown losses of 10q, 17q and 19 with additional 5q and 13q gains.[6]

Clinically subcutaneous panniculitis like T-cell lymphoma is characterized by indolent, slowly expanding, subcutaneous nodules and indefinite firm plaques involving the legs (71%), arms (62%), torso (56%), and/or face (25%).[4,7] Approximately 80% of patients have multiple nodules and/or plaque. Initially these lesions may be misdiagnosed as panniculitis. Occasionally patient present with more diffuse erythematous induration mimicking cellulitis. Ulceration is rare. Patients often have fever, chills, malaise, weight loss, night sweats. Systemic symptoms may occur particularly in those who develop Hemophagocytic syndrome consisting of fever, pancytopenia and hepato splenomegaly and may be associated with a rapidly progressive

downhill course and early relapse.[4,8] The hemophagocytic syndrome may occur secondary to lymphokine production by the malignant cells or may be related to the destruction of normal cells at subcutaneous tissue.[1] A literature review of 16 Japanese with SPTCL showed a higher incidence of B symptoms (81%) and hemophagocytic syndrome (45%) and a lower incidence of autoimmune disease (13%).[9]

Histologically there is a diffuse infiltrate restricted to and extending throughout the sub cutis without epidermotrophism.[4] subcutaneous infiltrate simulates a lobular panniculitis. Infiltrate contain a mixture of neoplastic pleomorphic cells of various sizes and macrophages. Rimming of individual fat cells by neoplastic T cells is helpful diagnostic feature.

The subcutaneous panniculitis like T cell lymphoma shows an alpha/beta+, CD3+, CD4-, CD8+, CD30-, CD56- immunophenotype.[8] They also express TIA-1+, granzyme-β+, and βF1+. The expression of β F1 (TCR α/β) by immunohistochemistry is a pivotal diagnostic marker for this entity. The neoplastic cells showed clonal T cell receptor gene rearrangements, although do not express specific genetic abnormalities and Epstein-Barr virus has not been identified.

Most of patients with subcutaneous panniculitis like T-cell lymphoma have an indolent course and recurrent episodes of eruption of subcutaneous nodules. The overall survival rate is about 80% and occasionally they resolve spontaneously. Multidrug combination chemotherapy is no longer used as first line therapy to treat the disease. Chemotherapy with cyclophosphamide, doxorubicin, vincristine, and prednisolone regimen showed overall remission rates of 50%. Immunosuppressive therapy can result in complete cure in patients with subcutaneous panniculitis like T cell lymphoma. Systemic corticosteroid in combination with cyclosporine A or methotrexate can be good strategy of treatment.[10-14]

Conclusion:

Subcutaneous panniculitis like T cell lymphoma is a rare entity with relatively difficult to diagnose due to the fact that clinical manifestations are similar to those of some cases of panniculitis. So, the final diagnosis should be made based on a combination of the patient's clinical manifestations, histopathological findings and immunophenotyping.

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