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CASE REPORT ON SUBCUTANEOUS PANNICULITIS;LIKE;T-CELL LYMPHOMA: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT Subcutaneous Panniculitis like T-cell lymphoma is one of rarest form of lymphoma with aggressive clinical behaviour characterized by primary involvement of subcutaneous adipose tissue without lymphnode involvement. SPTCL is difficult to diagnosed as its symptoms are non-specific and can be mimics with other conditions like benign panniculitis, eczema, dermatitis, psoriasis and cellulitis. It accounts for fewer than 1% of all extranodal non Hodking lymphomas. Here we reported a case of a young woman with Subcutaneous Panniculitis like T-cell lymphoma who achieved complete remission after chemotherapy.

KEYWORDS : SPTCL, Extranodal Non Hodking lymphoma

INTRODUCTION

SPTCL (Subcutaneous panniculitis-like T-cell lymphoma) is one of the rarest form of skin lymphoma that affects the subcutaneous adipose tissue and it has no lymph nodes involvement. [1] and it accounts for <%1 of all extranodal Non-Hodgkin's lymphomas (NHL) . It was first described in 1991 and was later recognized by WHO in 2001 [2]. Diagnosis of SPTCL is difficult at initial stage as symptoms mimic other conditions like benign panniculitis, dermatitis, psoriasis and other skin infections. Subcutaneous nodule, fever with or without chills, and weight loss are nonspecific clinical and systemic symptoms that are seldom linked with hepatomegaly , splenomegaly , mucosal ulcers, pleural/pericardial effusions, and rarely with hemophagocytosis syndrome (HPS) [3, 4]. The histologic features are lymphoid infiltration , primarily involving the interstitium of fat lobules that resembled lobular panniculitis without infiltration to papillary dermis and epidermis except in a some cases of cutaneous relapse. The disease process follows an waxing and waning pattern, with no constitutional symptoms and often spontaneous regression of skin lesions [5]. Here we reported the case of 30 year female who initially presented with subcutaneous nodules and was diagnosed as SPTCL on histological investigation. She was managed with chemotherapy and is on complete remission after treatment.

CASE REPORT

A 30 year lady presented with chief complaints of painless subcutaneous nodule at the back and buttocks associated with fever on and off since 3 months. The patient was initially evaluated and managed outside as case of panniculitis under Dematology department. However the patient doesnot respond to the treatment and then came to our center with chief complaints of increase in the size of subcutaneous nodule at the back and left thigh region . On examination multiple firm to hard ~1-5cm sized subcutaneous nodules were palpable over chest , back , buttocks and left thigh and dimpling sign present (Figure 1 {A}). Her skin biopsy from left thigh and back nodule show periadenexal and subcutaneous fat shows lobular lymphocytic infiltrate features consistent of Lupus panniculitis (Figure 2). The cells were positive for CD-3, CD-8, S-100, TCRβF1 and negative for CD-4, CD-30, CD-56, CD-20, and EBV on immunohistochemistry, with a Ki67 score of 70%, indicating Subcutaneous Panniculitis -like T cell lymphoma. Her initial LDH (lactate dehydrogenase) level was 470 IU/L. Radiologically imaging 18F-fluoro-2-deoxy-D-glucose (18F-FDG) positron emission tomography/computed tomography (PET/CT) scan showed FDG avid multiple ill defined lesion noted at subcutaneous region of back , gluteal and left lateral thigh with SUV max 6.8. She was initially started on oral methotrexate and prednisolone based chemotherapy however in view of waxing and waning pattern of disease she was later planned for intravenous chemotherapy. She received 3 cycles of Gemcitabine and cisplatin based chemotherapy . Post chemotherapy in view of stable disease the patient was planned for change in CHOP based chemotherapy . She received 4 cycle of chemotherapy with vincristine, adriamycin , cyclophosphamide and prednisolone. Her skin lesions were completely resolved after 2 cycle of chemotherapy (Figure 1 {B}) . Response assessment was done after completion of therapy which shows complete response on PET CT scan. After 17 months of treatment, she is now asymptomatic and in total remission.

DISCUSSION

Subcutaneous Panniculitis-like T-cell Lymphoma is one of the rarest form of T cell lymphoma, which was recognized in World Health Organization (WHO) classification as distinct entity in 2001 [1]. It's a type of cytotoxic T-cell lymphoma marked by subcutaneous infiltrates of variable-sized pleomorphic T cells and a high number of macrophages. It occurs in adults as well as in young children, and equal prevelance in both sexes. Multiple subcutaneous swellings and/or erythematous plaques or ulcerated skin nodules are the most prevalent dermal lesions, which affect the trunk, extremities, and face. It rarely associated with hemophagocytic syndrome, which indicate a rapidly progressive disease course [3].

As with any other type of lymphoma, radiological imaging is useful for diagnosing and for assessment of treatment response. Punch biopsy of the lesion of skin and subcutaneous tissue, immunohistochemical labelling, and molecular analysis are used to make the diagnosis.

Histologically showed a densed atypical of small- to medium- to large-sized lymphocyte infiltration and characteristically involving the fat lobules mimics a lobular panniculitis but typically sparing the septae and overlying dermis . Some cases show features of granuloma formation, which mimics of granulomatous panniculitis (4). About 20% of cases involve autoimmune disorders. Due to the similarities in clinical and histologic characteristics, lupus erythematosus panniculitis (LEP) is typically included in the differential diagnosis , which can be differentiated SPTCL by the lack of cytologically atypical T-cell (6).

Immunohistochemistry of SPTCL shows that cells are CD-2, CD-3, CD-7, CD-8, beta F1,and activated cytotoxic proteins like TIA-1 (T-cell intracellular antigen) are positive, while CD-4, CD-56, and CD-30 are negative [7]. Based on the phenotype of T-cell receptors and IHC staining, there are two forms of SPTCL: (i) TCRαβ , which has an indolent course and is generally CD8+ve , CD4-ve and CD56-ve and (ii) TCRγδ, which has a rapid clinical deterioration and is usually CD8-ve , CD4-ve and CD56+ve [8]. The γδ phenotype is usually associated with hemophagocytosis and associated with adverse prognosis, while α/β Tcells forming around 70% of cases with prolonged survival. 18F-fluoro-2-deoxy-D-glucose (18FDG) positron emission tomography/computed tomography (PET/CT) scan imaging investigations are useful for determining the number of lesions with aberrant uptake and assessing therapy response. Treatment options include topical, immunosuppressive, and cytotoxic medicines, which can be used in combination or alone, similar to how other cutaneous T-cell lymphomas are treated [7]. The most effective systemic treatment options were anthracycline-based chemotherapy regimens. Most of the time Anthracycline- or anthracenedione-based chemotherapy regimens are commonly employed, with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) or CHOP-like combinations (9). The preferred combination chemotherapy regimen is cyclophosphamide, doxorubicin, vincristine, and prednisolone, with overall remission rate of 50% (10). Autologous bone marrow transplantation has been benefited in rare circumstances. Tcell receptor (ASCT) patients had a 5-year survival rate of more than 80% (4). In present case report of young post 4 cycles of CHOP based chemotherapy is with no evidence of disease 17months after treatment completion with close followup.

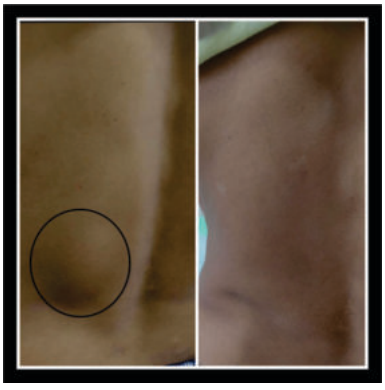

Figure 1 : (A) Pre Treatment Initial clinical presentation of patient with Slightly erythematous subcutaneous nodule on the back. (b) Post Treatment – Complete resolution of Nodule.

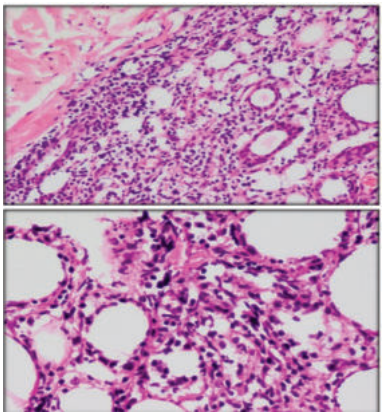

Figure 2: A punch biopsy section with hematoxylin and eosin stain shows infiltration of a-typical lymphoid cells encircling the fat lobules.

Conflicts of Interest : There are no conflicts of Interest

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