DOI: 10.7860/JCDR/2016/18458.7824

Case Report

Internal Medicine Section

Subcutaneous Panniculitis-Like T-Cell Lymphoma: A Rare Tumour

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ABSTRACT

Subcutaneous Panniculitis like T cell Lymphoma (SPTCL) is an uncommon variant and poorly differentiated type of cutaneous T cell lymphoma. Here we describe the case of a 19-year-old female who presented with swelling of left half of the face with no regional lymphadenopathy and hepatosplenomegaly which was initially misdiagnosed as a benign cutaneous condition by various practitioners. Histopathological examination revealed diffuse infiltration of subcutaneous plane by small to medium sized atypical lymphocytes. Immunohistochemistry showed CD3, CD8 and βF-1 positivity; CD20, CD56, Epstein Barr Virus (EBV) and TCR-δ negativity. Clinical profile, histopathology and immunohistochemical analysis yielded a diagnosis of SPTCL. Thus cases with atypical and nonresolving dermatological lesions should raise a suspicion of SPTCL as diagnosis against other benign conditions.

Keywords: Atypical lymphocytes, Cutaneous condition, Face, Non resolving

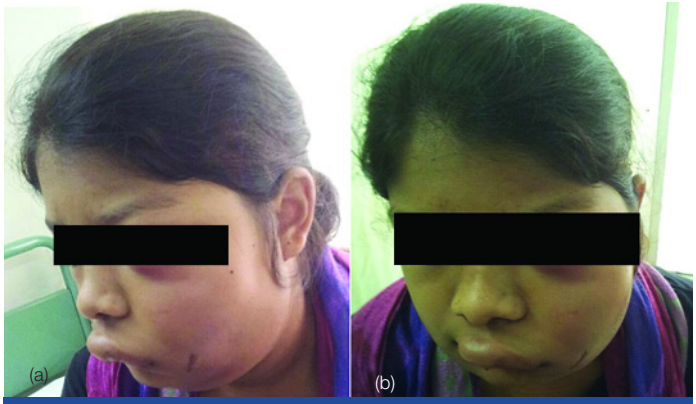
CASE REPORT

A 19-year-old previously healthy female, presented to us with 3 months history of diffuse swelling of left half of the face with recent increase in size of the swelling and fever for the past few weeks. There was no history of significant weight loss or other symptoms. She had history of numerous consultations with Dentists, Dermatologists, Otolaryngologists and Oro-maxillofacial specialists for the evaluation of the swelling. Several biopsies done from the swelling earlier at various centres were reported with varied diagnosis such as inflammatory/infectious panniculitis, spindle cell lipoma, granulomatous disease etc. She also received multiple courses of antibiotics but appreciated no improvement. Examination revealed swelling of left half of the face which was erythematous, indurated and tender [Table/Fig-1a&b]. There was no regional lymphadenopathy and hepatosplenomegaly.

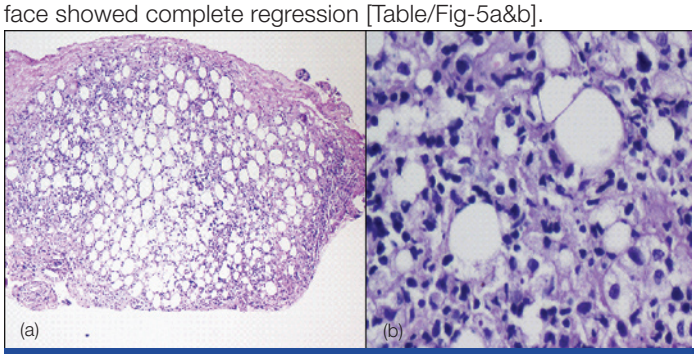
Her routine haematological and biochemical parameters were within normal limits. Ultrasound of the swelling was suggestive of diffuse skin and subcutaneous tissue involvement of the left maxillary, buccal, masseteric regions with hyperechoic lesion measuring 2.5x1.7cm. Deep biopsy showed lobules of adipocytes and muscle bundles infiltrated by small and medium sized lymphocytes showing atypical, hyperchromatic nuclei with foci of necrosis and karyorrhexis. Foci of fat necrosis with fibrosis and proliferating capillaries were also seen [Table/Fig-2a&b]. Immunohistochemistry showed CD3, CD8 and βF-1 positivity; CD20, CD56, Epstein Barr Virus (EBV) and TCR-δ negativity [Table/Fig-3a&b].

Hence a diagnosis of SPTCLα/β subtype was made based on the clinical presentation, histopathological features and immunohistochemistry. Whole body Positron Emission Tomography (PET) scan showed diffuse subcutaneous thickening and stranding with linear plaque like Fluorodeoxyglucose (FDG) uptake in left half of the face, left preseptal region with no regional lymph node involvement and hepatosplenomegaly [Table/Fig-4a&b]. Patient was staged as STAGE IE (WHO-EORTC classification).

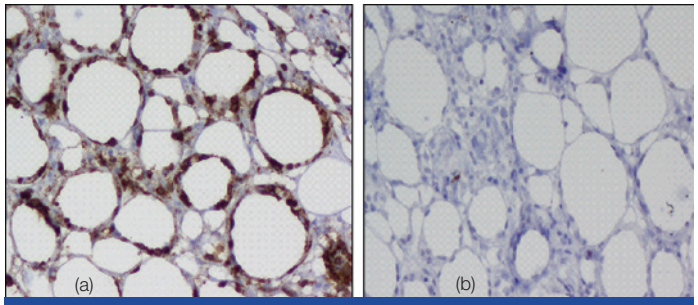
She was treated with Cyclophosphamide, Adriamycin, Vincristine, Prednisolone and Etoposide (CHOPE) regimen following which fever subsided and overall condition improved. By second cycle of chemotherapy patient reported remission of the existing lesion and there were no new lesions on follow-up. Patient received a total of 4 cycles of chemotherapy and clinically swelling over the face showed complete regression [Table/Fig-5a&b].



[Table/Fig-1a, b]: Showing swelling of left half of the face.



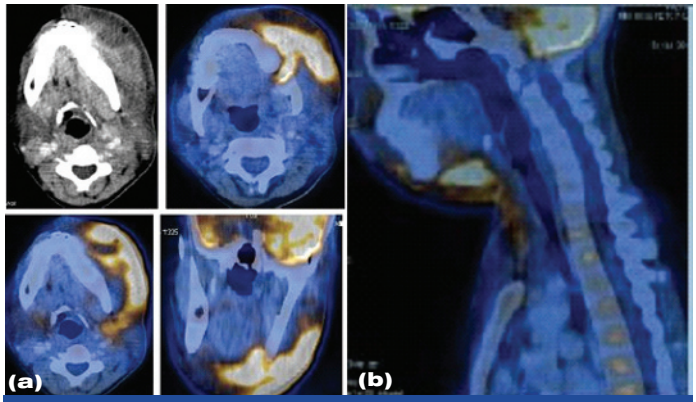
[Table/Fig-2a,b]: Photomicrograph showing fat planes infiltrated by atypical small to medium sized lymphocytes, bordering the adipocytes.



[Table/Fig-3a, b]: Atypical lymphocytes showing (a) CD 3 positivity and (b) CD 20 negativity on immunohistochemical staining.

DISCUSSSION

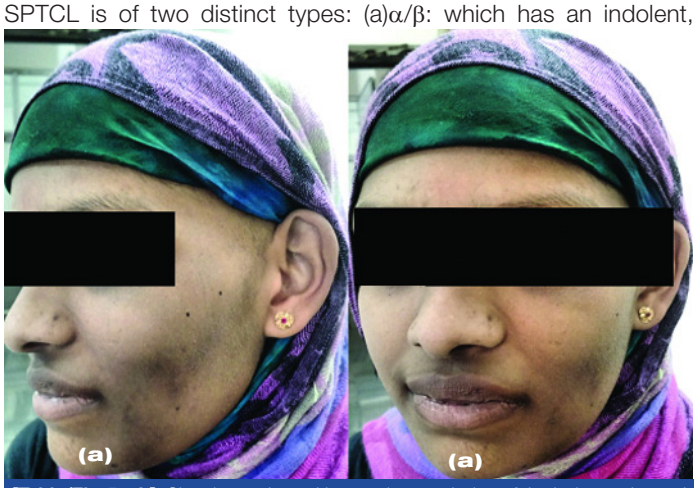
Subcutaneous Panniculitis like T-cell Lymphoma (SPTCL) is an uncommon type of cutaneous lymphoma initially described by Gonzalez in 1991 [1]. REAL (Revised European American Lymphoma) and EORTC (European Organization for Research



[Table/Fig-4a, b]: PET scan images showing FDG uptake in the left half of the face, left preseptal region extending caudally up to the suprasternal notch.

and Treatment of Cancer) classification of cutaneous tumours considered SPTCL as a provisional entity [2], which was subsequently considered as a distinct cutaneous lymphoma by WHO in 2001 [3].

Based on the T-Cell Receptor (TCR) and immunophenotype, SPTCL is of two distinct types: (a)α/β: which has an indolent,



[Table/Fig-5a, b]: Showing patient with complete remission of the lesion at the end of fourth cycle of chemotherapy.

less aggressive course; CD8+, CD4- and CD56- (b) γ/δ: has a rapid and fatal course due to associated Haemophagocytosis Syndrome(HPS) and is usually CD4-, CD8- and CD56+. Usually the patients present with plaques and subcutaneous nodules involving the extremities without lymph node involvement and diagnosis may become difficult as the symptoms mimic conditions like eczema, cellulitis, dermatitis, benign panniculitis etc. Clinical course of the disease is mostly indolent but rapid progression is not uncommon. It is less commonly associated with hepatosplenomegaly, haemophagocytosis syndrome, cytopenias and these associations indicates poor prognosis. The overall 5 year survival rate in SPTCL α/β is 82% whereas in SPTCL γ/δ it is as low as 11% [3]. It is mostly malignancy of young, which usually presents in second decade of life. Go and Wester study showed that 75% of the patients were aged between 18 -60 years which is consistent with this case [4]. Patients with SPTCL usually present at dermatology clinic with plaques and nodules involving the extremities [5]. This patient had history of numerous visits to various centres, where it was misdiagnosed as a benign skin disease.

The clinical presentation was unique in this case, as the patient presented with diffuse swelling of left half of the face in contrast to the previously described cases. Kosari et al. and Cassis TB et al., have also reported cases wherein face was the initial site of involvement [6,7]. This patient had no lymph node involvement, hepatosplenomegaly and haemophagocytosis syndrome indicating the less aggressive form of the disease. SPTCL shows neoplastic lymphoid cell infiltration on histology, which exhibit atypical features like hyperchromatic and angulated nuclei with hazy cell borders. There may be benign histiocytes, neutrophils and plasma cells, mimicking more common condition like benign panniculitis [8]. Rimming of the fat cells with atypical lymphocytes, focal areas of fat necrosis, apoptotic cells and debris from karyorrhexis are also seen in SPTCL. Immunohistochemistry plays a critical role in the diagnosis [9-11]. Positron Emission Tomography [PET] scan is used for the diagnosis, staging and also for monitoring the treatment response [12]. Different treatment regimens have been mentioned but CHOPE like regimen are the most commonly used [5] and even the patient in this report responded well to CHOPE based regimen with complete remission by second course of chemotherapy.

CONCLUSION

Subcutaneous panniculitis like T cell lymphoma is an infrequent disease which is often missed or misdiagnosed by clinicians as well as pathologists for other benign and commoner conditions. This case emphasizes the fact that high degree of suspicion and expertise is important for diagnosing such uncommon malignancy. Hence early diagnosis of such rarer malignancies helps in appropriate treatment which ultimately leads to better chance of survival and reduces the risk of fatality.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Dec 22, 2015**
Date of Peer Review: **Mar 15, 2016**
Date of Acceptance: **Mar 26, 2016**
Date of Publishing: **May 01, 2016**