CASE REPORT

Subcutaneous Panniculitis-like T-cell Lymphoma: A Case Report of A Solved Conundrum

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ABSTRACT

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare primary cutaneous lymphoma which often be very complexed to be diagnosed. Here we reported a case of a 38-year-old lady with wax and wane course of left facial swelling associated with prolonged pyrexia of unknown origin, lethargy and prominent weight loss. Multiple investigations were conducted to reach to the conclusive diagnosis. Histopathological showed infiltration of neoplastic lymphoid cells within the adipocyte clusters with hyperchromatic nuclei, small nucleoli and scant cytoplasm. Immunohistochemical analysis were immunoreactive to CD3, CD8 & TIA and negative to CD20, CD4 & CD56. Based on clinical findings, histopathology report and immunohistochemical analysis, the patient diagnosed with SPTCL. Hence, any suspicious maxillofacial swelling without obvious sign of infection and trauma shall be investigated until a final diagnosis is reached.

Keywords
Lymphoma, Facial, Swelling, Subcutaneous, Panniculitis.

INTRODUCTION

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare primary cutaneous lymphoma which is often very complex to diagnose. We report here a case of a 38-year-old lady with wax and wane course of left facial swelling associated with prolonged pyrexia of unknown origin, lethargy and prominent weight loss. Multiple investigations were conducted to reach the conclusive diagnosis. Histopathological findings showed infiltration of neoplastic lymphoid cells within the adipocyte clusters with hyperchromatic nuclei, small nucleoli and scant cytoplasm.

Immunohistochemical analysis were immunoreactive to CD3, CD8 & TIA and negative to CD20, CD4 & CD56. Based on clinical findings, histopathology report and immunohistochemical analysis, the patient was diagnosed with SPTCL. Hence, any suspicious maxillofacial swelling without obvious sign of infection and trauma should be investigated until a final diagnosis is reached. The patient was started with immunosuppressive therapy – mycophenolate mofetil. She is still undergoing mycophenolate mofetil for six weeks with signs of improvement noted. A review after 12 months showed no signs of recurrence of the disease.

CASE DESCRIPTION

A 38 years old Malay female was referred to us at Oral & Maxillofacial Surgery Unit, SASMEC @IIUM after multiple tests were done to diagnose her peculiar clinical condition which seemed to be inconclusive. This patient presented with multiple intermittent swelling episodes at back of her neck and pubic region which regressed on their own.

This was followed by swelling on her facial region which was painless with normal skin colour, which increased and decreased in size intermittently on a daily basis. Her condition was also associated with intermittent episodes of pyrexia, lethargy and there was prominent weight loss. The patient denied any history of insect bite or toothache prior to the swelling episode. She also had history of similar swelling on the right side of her face one month earlier which subsequently resolved with steroid.
Our patient had experienced prolonged pyrexia of unknown origin since February 2020 and was under investigation for plasma cell dyscrasia. Patient didn’t present with any neurological symptoms. Full blood picture showed normochromic normocytic anaemia with moderate anisopoikilocytosis and leucopenia with occasional reactive lymphocytes. Iron study was done and showed low serum iron and Total Iron Binding Capacity, but high serum ferritin. Bone marrow aspiration and trephine biopsy was done in July 2020 showing normocellularity with all stages maturation. Serum free kappa/lambda ratio was within normal range (1.064). There were raised serum IgG and IgA. Serum protein electrophoresis showed normal protein levels. Urine protein electrophoresis, however, showed higher protein level.

There were persistently increased inflammatory markers (ESR, CRP & Procalcitonin), CK, LDH and liver enzymes values throughout admission periods. The APTT value was raised consistently. Autoimmune markers (ANA, ANCA, C3, C4, anti-smooth muscle antibody) showed negative results. Tumour markers (AFP, CEA, Ca 125, Ca 19-9) showed mild elevation. The triglyceride level was on normal range. Blood culture, urine culture, BFMP, leptoseroology, Mantoux test and viral screening were done to rule out any infection and came out as negative. Clinically, there was no evident of infection from extraoral and intraoral regions. Orthopantomogram (OPG) was taken but no obvious sign of pathology noted. Paranasal sinus CT scan showed diffuse soft tissue thickening involving the left periorbital, maxillary and mandibular with no suspicious features. No focal bony lesion or bone destruction evidenced. An incisional biopsy was performed via intraoral approach where soft tissue from lateral side of the nose and orbicularis oris muscle were taken and sent for histopathological examination (HPE). Histologically, there was infiltration of neoplastic lymphoid cells within the subcutaneous adipocytes (Figure 2). The tumour cells exhibited hyperchromatic nuclei with small nucleoli and scant cytoplasm.

The background showed many reactive histiocytes, necrotic adipocytes and apoptotic bodies (Figure 3). Mitosis is also evident. These neoplastic cells were immunoreactive to CD3, CD8, TIA and negative to CD20, CD4 and CD56 (Figure 4). They showed high proliferative index about 80% as expressed by ki67. Subsequently, a diagnosis of subcutaneous panniculitis-like T-cell lymphoma (SPTCL) was made. The more aggressive type, panniculitis-like feature of cutaneous gamma-delta T-cell lymphoma was excluded by the negative CD56 staining.
are treated with various options: ranging from topical to immunosuppressive agents all the way to cytotoxic agents, in combination or alone. Bhojaraja et al. for example, reported a case of SPTCL with left facial swelling treated with CHOPE (Cyclophosphamide, Doxorubicin, Vincristine, Prednisolone & Etoposide) based regime. In our case, the patient was found to fit all criteria for SPTCL and was started with immunosuppressive therapy – mycophenolate mofetil, similar to a case reported by Heyman and Beaven. They observed significant reduction in the size and number of cutaneous lesions, few side effects, and an improvement in overall quality of life, which is ongoing after 12 months of therapy. There is no specific treatment guideline for SPTCL available in literature.

Diagnosing SPTCL can be quite a challenge and therefore detailed history taking, investigations and multi-disciplinary management is recommended. Primary cutaneous lymphomas (CL) are extranodal non-Hodgkin lymphoma which occur in the skin without extracutaneous involvement during diagnosis and staging period. CLs were initially considered as cutaneous manifestation of systemic lymphoma. Primary CLs differ in its biological behaviour and prognosis from histologically or phenotypically similar nodal and extranodal lymphomas. Due to this reason, CLs are now recognized as different group of lymphomas. Classification of CLs have undergone dynamic changes since it was first encountered. Presently, the 2018 update of World Health Organization-European Organization for Research and Treatment of Cancer (WHO/EORTC) classification is the gold standard for diagnosis and treatment planning.

SPTCL was initially subtyped into its α/β and γ/δ phenotypes. In the revision of WHO/EORTC classification and WHO classification (4th edition, 2008), this term is by definition restricted to cases expressing a T-cell receptor (TCR) α/β phenotype. The γ/δ phenotype is now classified under rare subtype of primary cutaneous peripheral T-cell lymphoma as primary cutaneous γ/δ T-cell lymphoma (Cγδ-TCR). Phenotypically, SPTCL expresses CD8 and cytotoxic proteins while Cγδ-TCR expresses CD56, CD2 and CD3 but are negative for CD4 and CD8. SPTCL is more indolent with 5-year disease specific survival (DSS) of 87% while Cγδ-TCR is more aggressive with 5-year DSS of 11%. Patients with SPTCL.

Therefore, Alsomali et al did a systematic review of nine articles reporting 15 cases of SPTCL consisting various treatment modality and suggested for lymphoma societies to publish a guideline regarding this. Even though SPTCL are typically described with subcutaneous nodules or plaques at upper and lower extremities and trunk, there are few cases reporting SPTCL with facial swelling in the literature similar to this case. The most commonly involved areas include the legs (71%), arms (62%), trunk (56%), and/or face (25%).

Figure 4: The atypical lymphocytes are immunoreactive to CD3, CD8 and TIA, but negative for CD56 (200x magnification).

DISCUSSION

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