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Case Report

A rare case of peripheral T cell lymphoma, not otherwise specified with cutaneous involvement and diffuse systemic metastasis

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ABSTRACT

Peripheral T-cell lymphoma (PTCL), a subdivision of T-cell non-Hodgkin lymphomas (NHLs), is rare and different from the more common cutaneous T-cell lymphomas. PTCL is a diverse group of disorders and carries a poor prognosis. Peripheral T cell lymphoma not otherwise specified (PTCL-NOS) is the most common and aggressive disorder out of the group, which involves the lymph nodes followed by the skin, liver, and gastrointestinal tract. We hereby report a case of a 60 years old female who presented to us with complaints of multiple painful dark-coloured nodular lesions and indurated plaques over bilateral upper limbs, lower limbs and trunk, for 3 months. After detail laboratory investigations, histopathological and immunohistochemistry which was positive for CD30 antigen we confirmed the diagnosis as PTCL-NOS. Radiological imaging showed involvement of gastrointestinal tract, adrenal gland and vertebrae. After thorough workup the patient was started on palliative radiotherapy, unfortunately she succumbed to death. In conclusion, PTCL-NOS with CD30 positivity is rare and aggressive with a poor prognosis, it is important to identify such cases and work in conjugation with an oncologist for proper management.

Keywords: Lymphoma, Non-Hodgkin, Immunohistochemistry, Cutaneous lymphoma

INTRODUCTION

Primary cutaneous lymphomas are a rare group of Non-Hodgkin lymphomas that include a heterogenous group of cutaneous T-cell lymphomas (CTCLs) and cutaneous B-cell lymphomas (CBCLs).[1] The basis for making a diagnosis is a clinico-pathological correlation, including the use of several immunohistochemical markers and molecular biological methods.[5]

CASE REPORT

A 60 years old female presented with complaints of painful dark coloured lesions over bilateral upper, lower limbs and trunk. The lesions started as light coloured patches over the right forearm and over a span of 3 months, spread to involve the upper and lower limbs and trunk. Cutaneous examination showed multiple hyperpigmented, firm to hard infiltrative nodules over the buttocks and lower limbs. Multiple hyperpigmented indurated plaques with xerotic surface were present over forearms, trunk, and thighs.

General examination was within normal limits. Extensive workup was done to rule out malignant and infective causes. Laboratory investigation showed raised ESR (65 mm/hr), low haemoglobin (8.2 grams/dl), elevated serum LDH (667 U/l) and uric acid (9.1 mg/dl). Buffy coat was negative for atypical lymphocytes. Pan CT scan showed multiple subcutaneous and left breast nodules and an enhancing lesion in the left adrenal gland. PET scan showed multiple disseminated cutaneous lesions as well as visceral lesions involving the mesentery, duodenum,

stomach, left adrenal gland and body of 3rd lumbar vertebrae which were likely to be lymphomatous deposits. Bone marrow biopsy was normal. Histopathological examination from nodular lesions showed irregular epidermal hyperplasia, nodular and dense diffuse infiltrate with bottom heavy appearance. Higher magnification showed epidermotropism and atypical mitoses. Immunohistochemistry was positive for CD3, CD4 and CD30 lymphocyte markers.

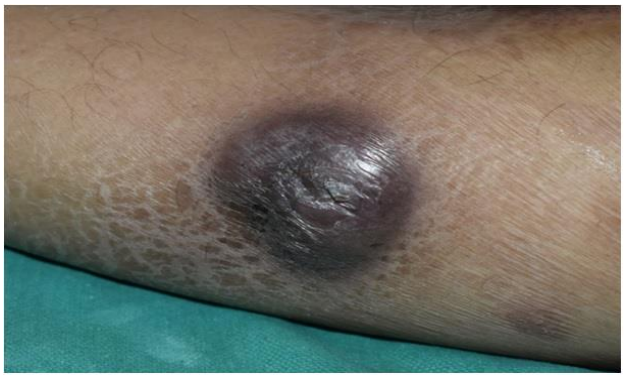

Figure 1: Hyperpigmented, tender, hard to firm nodules over lower limbs.

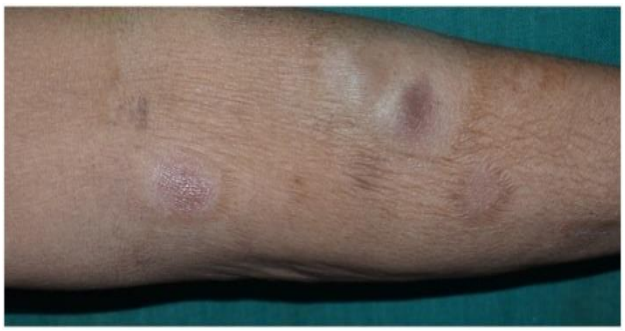

Figure 2: Multiple erythematous-hyperpigmented indurated plaques with xerotic surface over forearms.

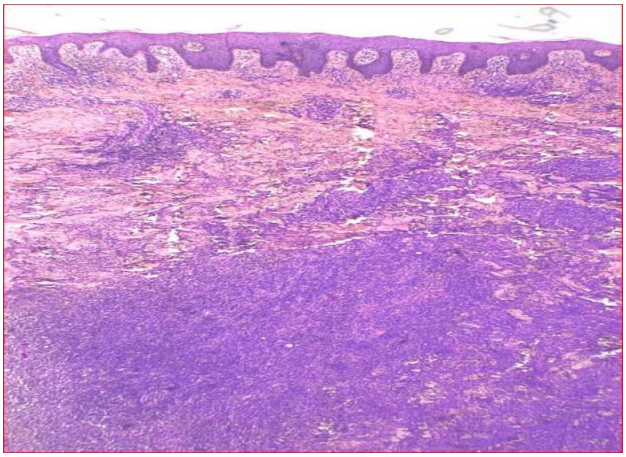

Figure 3: Histopathology showing irregular epidermal hyperplasia, nodular and dense diffuse infiltrate with bottom heavy appearance, lymphocytes in epidermi – epidermotropism.

Figure 4: Immunohistochemistry- the lymphoid cells are positive for cd3 with retained cd7 expression and show mild preponderance for cd4 over cd8. CD30 and CD56 are negative. MIB 1 labelling index <5%.

Thus, based on clinical, histopathological, immunohistochemical and radiological findings, a diagnosis of PTCL-NOS was made and the patient was started on palliative radiotherapy. Unfortunately, she succumbed to death due to sepsis.

DISCUSSION

PTCL, a subdivision of T-cell NHLs, is rare and different from the more common cutaneous T-cell lymphomas PTCL is a diverse group of disorders, it has 29 different subtypes and carries a poor prognosis.[1]

PTCL-NOS is the most common subtype of PTCL affecting adults in their 5th and 6th decade of life and usually affects males more than females.[3] It can be nodal and/or extra nodal; the extra nodal sites being the liver, bone marrow, gastrointestinal tract, and skin.[2] In our patient the skin and gastrointestinal tract was involved. Histopathology of PTCL-NOS shows mononuclear infiltrate of small to medium sized lymphocytes with few atypical cells.[4]

Phenotypically it lacks expression of T cell markers like CD5, CD7 and may express CD3, CD4 and/or CD8. PTCL and anaplastic large cell lymphoma-anaplastic lymphoma kinase negative (ALCL-ALK) both express CD30 antigen, but later has different morphology and expresses epithelial membrane antigen.[5] In our case, on immunohistochemistry small lymphoid cells were positive for CD3, CD4, CD30 and negative for CD5, CD7, CD8, CD20, PAX5 and ALK1.

Prognosis of PTCL is not as good as aggressive B cell lymphoma. Factors such as high age, raised serum LDH levels, bone marrow involvement cause poor prognosis. Biologic factors like expression of ALK1, TCR BF, NFKB, CCR3 have a more favourable prognosis than those which express p53, Ki67, Bcl-2, Bcl-x and CD30.[1] It has a 16-36% 5-year disease specific survival rate.[6] In our

case, high age, raised LDH levels, CD30 positivity were bad prognostic factor and she did not possess any risk for malignancy other than her age.

Treatment of PTCL-NOS depends on the stage and type; it includes chemotherapy with CHOP (cyclophosphamide, doxorubicin, vincristine, prednisolone) or ECHOP (etoposide, cyclophosphamide, doxorubicin, vincristine, prednisolone), immunotherapy, systemic steroids, stem cell transplantation, targeted therapy and radiation.[7] Once a diagnosis of PTCL is made, further tests need to be performed, to see the involvement of different organs and for the staging of the disease, such as positron emission tomography, computed tomography, bone marrow biopsy, lumbar puncture etc. Our patient was treated with palliative radiotherapy as she was in an advanced stage. She noticed ulcers over the nodular lesion and unfortunately succumbed to sepsis due to secondary bacterial infection.

CONCLUSION

Cutaneous T-cell lymphomas are a rare group of disorders out of which PTCL-NOS with CD 30 positivity is even rarer. It is important to be aware of the signs and symptoms of this disorder and thus have a clinical suspicion so that one can diagnose it appropriately. Since this has an aggressive clinical course and a poor prognosis, we need work up such cases quickly so that appropriate treatment can be started soon. As dermatologists, we need to work in conjunction with an oncologist as part of the team so that appropriate patient management involving chemotherapy and radiotherapy can be instituted.

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Ethical approval: Not required

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