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Case Report

A Rare Case of Angioimmunoblastic T-Cell Lymphoma with Pleomorphic Cutaneous Manifestations

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ABSTRACT

Angioimmunoblastic T-cell lymphoma is a type of mature T-cell lymphoma that is characterised by a range of symptoms including fever, night sweats, weight loss, swollen lymph nodes, enlarged liver and spleen, high white blood cell count, immune system dysfunction, fluid accumulation in the chest and abdomen and edema. The skin may also be affected, with various types of lesions and tumours such as maculopapular, urticarial, vesicular and nodular. Diagnosis requires a lymph node biopsy and immunohistochemistry, as the histological changes are typically subtle. This case is being presented to increase awareness of this rare and unique lymphoma, which can have a variety of skin symptoms and modest abnormalities that can make diagnosis difficult in clinical practice. To properly diagnose and stage the disease, additional tests such as histological assessment and immunohistochemistry are necessary.

Keywords: Peripheral T Cell Lymphoma, Cutaneous T cell Lymphoma, Pleomorphic cutaneous lesion

INTRODUCTION

Angioimmunoblastic T-cell lymphoma (AITL) is a mature T-cell lymphoma characterised by symptoms such as fever, night sweats, weight loss, swollen lymph nodes, enlarged liver and spleen, elevated white blood cell count, impaired immune system function, fluid accumulation in the pleural and peritoneal cavities and oedema.[1-3] Cutaneous manifestations may include maculopapular lesions, urticarial lesions, vesicular lesions and nodular tumours.

CASE REPORT

A 67-year-old man presented with asymptomatic red raised lesions on his trunk and neck, as well as axillary lymphadenopathy. A nodule was present on his trunk, and maculopapular lesions were observed on his face, chest and neck, as well as purpuric papules and plaques on his leg and thigh. In addition, he developed urticarial plaques and vesicular lesions [Figure 1]. His complete blood count revealed anaemia and eosinophilic neutrophilic leucocytosis. Abdominal and pelvic ultrasound findings were normal. Computed tomography of the neck revealed enlarged adenoids, tonsils and numerous enlarged bilateral cervical, left axillary and upper mediastinal lymph nodes. A skin biopsy conducted on the maculopapular and purpuric lesion and axillary lymph node biopsy was taken. The skin biopsy of the purpuric lesion revealed moderately thick superficial and mid-perivascular infiltration of neutrophils, eosinophils and lymphocytes. A few large atypical cells were observed in the perivascular and interstitial zones. The arteries were dilated with intravascular

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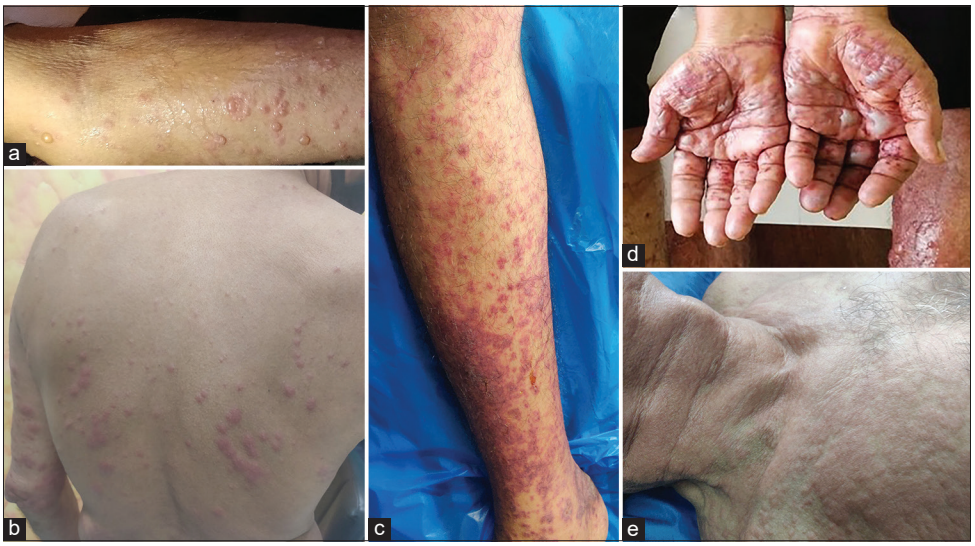

Figure 1: (a) Cutaneous lesion nodular, (b) macular papular, (c) purpuric, (d) vesicular and (e) urticarial.

and perivascular fibrin deposition, leucocytoclasia and red blood cell extravasation, indicating leucocytoclastic vasculitis. A biopsy of a maculopapular lesion revealed an extremely sparse superficial and mid-perivascular and periappendageal infiltration of eosinophils, few neutrophils, lymphocytes and occasionally large atypical lymphocytes in the upper dermis [Figure 2]. The results of the biopsy of the axillary lymph node revealed the presence of immunoblasts, eosinophils, plasma cells and cells with a mummified Reed-Sternberg appearance [Figure 3]. The immunohistochemistry of the lymph node indicated the presence of a malignant cell population that was positive for CD3, CD4, CD5, CD10 and BCL6. The immunoblastic cells and those with an RS-like shape expressed leucocyte common antigen, CD20, CD30, BCL6 and Epstein–Barr virus (EBV), but not CD15. The CD21 and CD23 showed an enlarged follicular dendritic cell network encircling the high endothelial venules and neoplastic cells [Figure 4]. The diverse range of cutaneous presentations with axillary lymphadenopathy, along with the lesional skin histopathology, revealed atypical cells along with leucocytoclastic vasculitis. In addition, the axillary lymph node biopsy revealed immunoblasts with morphological features resembling Reed-Sternberg and a malignant cell population that tested positive for CD3, CD4, CD5, CD10, Bcl6, EBV, CD21 and CD23. Consequently, the patient was diagnosed with AITL. The patient was referred to an oncologist for further evaluation. For the cutaneous signs, the patient was prescribed prednisolone 30 mg tablets and hydroxyzine 10 mg tablets for symptomatic relief, as well as a topical steroid cream.

DISCUSSION

AITL is a form of cancer that affects mature T-cells and accounts for 15–20% of peripheral T-cell lymphomas and 1–2% of non-Hodgkin lymphomas. It is most commonly found in elderly individuals with no clear gender preference.[4] Although cutaneous lesions are a common occurrence, occurring in up to 50% of patients, they can vary and resemble viral exanthems or medication rashes. The associated pruritus mainly presents as generalised morbilliform or maculopapular eruptions on the trunk, but less commonly, patients exhibit pleomorphic rashes with nodules, urticaria, petechiae, purpura and erythroderma. Rare cutaneous symptoms also include necrotic purpura, polyarthritis, gingival ulceration, erythematous plaques (sometimes annular), toxic epidermal necrolysis, hemorrhagic or necrotic nodules, vesicles and papulovesicular (prurigo-like) lesions.[5] The histological findings from the skin biopsies were subtle.[4] The presence of skin lesions is linked to a poor prognosis, similar to other non-cutaneous peripheral T-cell lymphomas. AITL can be treated with chemotherapy, stem cell transplantation, radiation and molecular targeted therapy; however, the outcomes are often disappointing, with a 30% 5-year survival rate.[6] Common markers employed in AITL diagnosis include CD20 and PAX5, which aid in identifying B-cell follicles and assessing the distribution, proportion and cytological features of B-cells. B immunoblasts and Hodgkin and Reed/Sternberg (HRS)-like cells tested positive for these markers. CD3 is an important marker used to evaluate T-cell distribution, proportion and cytological features, as well as to identify T-cells with atypia and altered expression of pan-T-cell antigens. Loss of CD3 and CD7, along with diminished CD2 and CD5 expression, is common in AITL. CD21 and CD23 are used to identify the FDC meshworks and extrafollicular proliferation, with CD21 being more sensitive than CD23. CD10 and BCL6 are employed to highlight reactive germinal centres and identify atypical T-cells with a T follicular helper (TFH) phenotype, serving as important but less sensitive markers for AITL

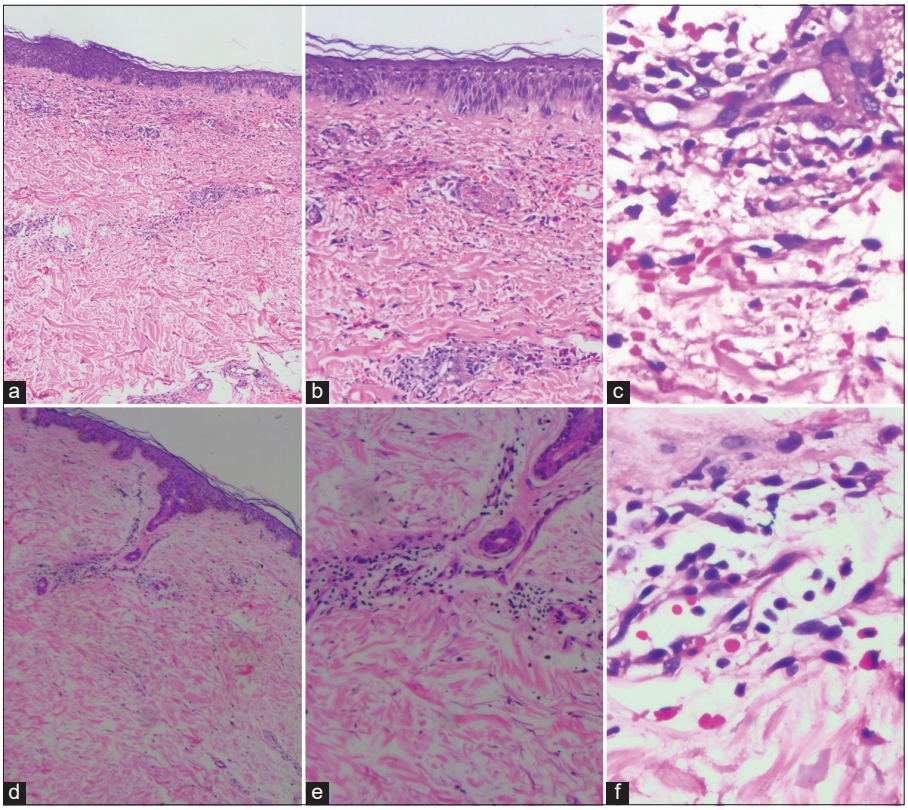

Figure 2: (a and b) biopsy from purpuric lesion H/E 10 × moderately dense perivascular infiltrate of neutrophils, eosinophils, lymphocytes. (c) H/E 40 × Few large atypical cells are seen in the perivascular and interstitial areas. There is dilatation of the superficial blood vessels with intravascular and perivascular fibrin deposition and leucocytoclasia at places suggestive of vasculitis. (d and e) A biopsy of a maculopapular lesion revealed an exceedingly sparse superficial and mid-perivascular and periappendageal infiltrate. (f) H/E 40 × infiltration of eosinophils, few neutrophils, lymphocytes, and occasionally large atypical lymphocytes in the upper dermis. H/E: Haematoxylin and eosin.

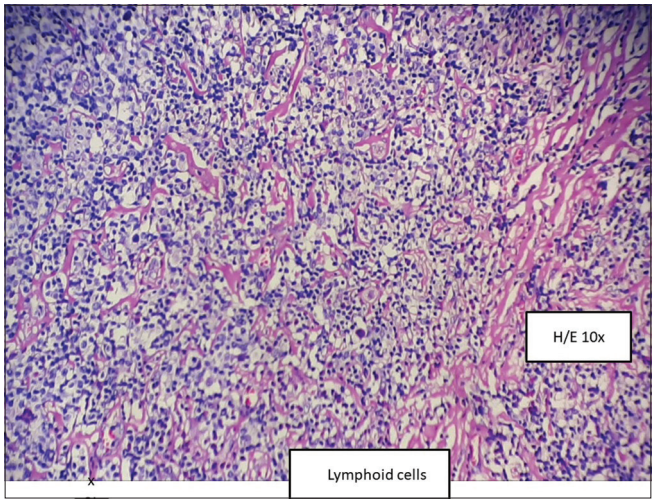

Figure 3: Histopathology of lymph node showing complete loss of nodal architecture. No germinal centres are seen. There is predominantly paracortical expansion comprising polymorphic medium-sized cells with clear/pale cytoplasm, distinct cell membranes and minimal cytologic atypia. The neoplastic cells are in clusters, typically admixed with variable numbers of polymorphous population of non-neoplastic cells comprising small reactive lymphocytes, eosinophils, plasma cells and histiocytes. H/E: Haematoxylin and eosin.

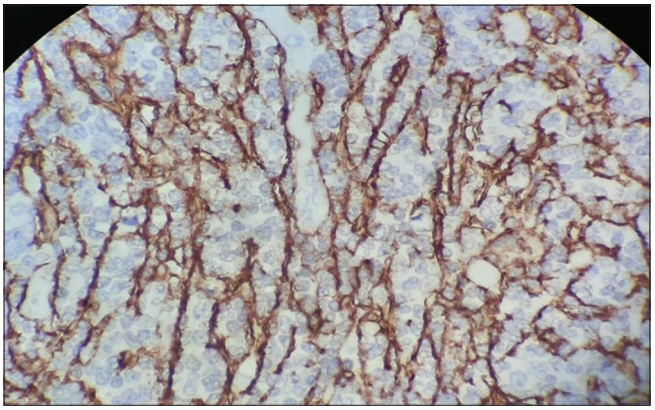

Figure 4: CD21 ×40 – Highlighting expanded follicular dendritic network. CD21: Cluster of differentiation 21.

diagnosis. programmed cell death protein 1 (PD1), inducible T-cell COStimulator (ICOS) and C-X-C Motif Chemokine Ligand 13 (CXCL13) help identify atypical T-cells with a TFH phenotype, but have variable sensitivity and specificity. CD30 is used to highlight B immunoblasts and HRS-like cells and may identify a subset of atypical T-cells. CD30 was employed

to highlight B immunoblasts and HRS-like cells, and may identify a subset of atypical T-cells. In approximately 20–30% of AITL cases, CD30+ atypical T-cells are present. κ and λ were used to evaluate light chain restriction in plasma cells, while EBV *in situ* hybridisation was used to identify EBV-positive B blasts. Collectively, these markers were used to assess the composition and characteristics of both B and T-cells in the infiltrate, aiding in the diagnosis of AITL.[7] The purpose of this case was to raise awareness among dermatologists of this rare and unusual lymphoma with varied cutaneous manifestations and subtle histological findings that can pose a diagnostic challenge in clinical practice. Accurate diagnosis and staging require complete investigation, including histopathological evaluation, lymph node biopsy and immunohistochemistry.

CONCLUSION

This case study highlights the diagnostic challenges posed by Angioimmunoblastic T-cell lymphoma (AITL) due to its diverse clinical presentations and subtle histological features. AITL, although rare, should be considered in the differential diagnosis of patients presenting with cutaneous lesions accompanied by lymphadenopathy. Clinicians should maintain a high index of suspicion and conduct thorough investigations, including skin biopsies and lymph node biopsies, supplemented by immunohistochemical analysis, to achieve an accurate diagnosis.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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