

Case Report

Skin manifestation of angioimmunoblastic T-cell lymphoma mimicking Hansen's disease on histology

Anju Shukla^{1*}, Sunil Sharad Dabadghao², Surbhi Gupta¹, Priyanka Jain¹

¹Department of Lab Medicine, Sahara Hospital, Lucknow, UP, India

²Department of Clinical Hematology and Bone Marrow Transplant, Sahara Hospital, Lucknow, UP, India

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*Correspondence:

Dr. Anju Shukla,

E-mail: anjushukla2012@gmail.com

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ABSTRACT

Angioimmunoblastic T-cell lymphoma (AITL) is an uncommon form of peripheral T-cell lymphoma now put under the category of nodal T-cell lymphoma with follicular T helper phenotype. 50% cases of AITL have varied cutaneous manifestations including macules, papules, petechiae, purpura, nodules, non-specific rash and urticaria. Herein we present a case of AITL initially presented as maculopapular rash which on histology was mimicking as Hansen's disease with perineural and perivascular inflammation; however, an infective organism could not be demonstrated. Later on, a delayed diagnosis was made on lymph node biopsy and immunohistochemistry and patient responded well with chemotherapy.

Keywords: Angioimmunoblastic T-cell lymphoma, Cutaneous manifestation, Hansen's disease

INTRODUCTION

Angioimmunoblastic T-cell lymphoma (AITL) long considered as angioimmunoblastic lymphadenopathy with dysproteinemia, a preneoplastic disorder is now considered as a neoplasm of mature T follicular helper cells (TFH) characterized by systemic disease, polymorphous infiltrate involving lymph nodes, proliferation of high endothelial venules (HEVs) and follicular dendritic cells (FDCs) in world health organization classification of haematopoietic and lymphoid tissues, revised 4th edition 2017.¹ AITL is the most common subtype of peripheral T cell lymphoma (PTCL) account for 15-20% of non-cutaneous T cell lymphoma and represents only 1-2% of all non-Hodgkin's lymphoma (NHL).² AITL is the disease of middle aged and elderly and patients usually presents late with B- symptom, generalized lymphadenopathy, modest hepatomegaly, and skin rash. Skin rashes are seen in 20-

50% of AITL patients ranging from urticarial lesion to nodular tumors.³ We present a case of AITL with skin involvement which on histology was mimicking as Hansen's disease.

CASE REPORT

A 42 years old female presented with history of generalized weakness, intermittent fever, weight loss, skin rash and pain in abdomen. On examination there was no mucocutaneous bleed, rashes were seen on face, forearm and back Figure 1 (a and b) with mild splenomegaly, and lymphadenopathy. Bony tenderness was absent. At the time of admission her hemoglobin was 8.7 g/dl, total leucocyte count was 5330/cu mm with 73% polymorphs. Platelet count was 71000/cu mm. Liver function test (LFT) showed normal level of transaminases with reduced serum albumin. Serum creatinine was within normal limit and viral markers were negative.

ANA, ANCA were negative and serum complement levels were in normal limit. Direct Coomb's test and serum immunoglobulin levels were normal. Computerized tomography scan showed hepatosplenomegaly with multiple small retroperitoneal and mesenteric lymph nodes. Patient underwent a skin biopsy which showed periadnexal, perivascular, perineural lymphohistiocytic infiltrate Figure 1 (c-e). On Wade-Fite's acid fast stain no lepra bacilli was identified, and on ZN stain no AFB was seen. When no conclusion was drawn on skin biopsy patient was subjected to guided lymph node biopsy. Hematoxylin and eosin (H and E) stained sections of lymph node revealed diffuse replacement of lymph node architecture (pattern III) with marked proliferation of arborizing high endothelial venules (HEV), polymorphic infiltrate composed of small to medium sized lymphocytes, plasma cells, immunoblasts and abundant eosinophils Figure 2 (b and c). Normal germinal centres were absent and mitotic activity was frequent. On Immunohistochemistry majority of cells expressed CD3 (Figure 2d), CD5 (Figure 2e), and CD10. BCL-6 was negative and KI-67 was 20-30% (Figure 2f). CD20 was positive in interspersed B cells (Figure 2c) and perinodal area.

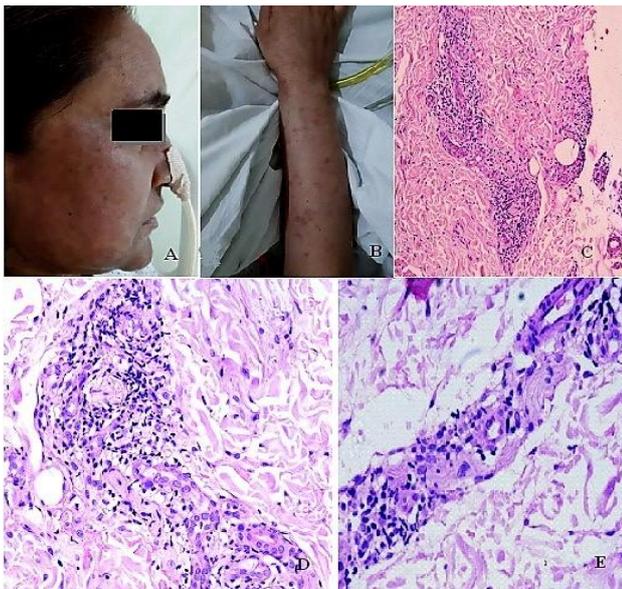


Figure 1: (a) Maculopapular rashes on face, (b) on forearm, (c) tissue section-skin biopsy showing perivascular, periadnexal and perineural infiltrate (H and E, X10), (d) lymphohistiocytic infiltrate with karyorrhectic debris (H and E, X40), and (e) perineural and intraneural infiltrate (H and E, X40).

Based on histomorphology and immunohistochemistry a diagnosis of angioimmunoblastic T-cell lymphoma was made and patient was given cyclophosphamide, vincristine, solumedral with premedication and transfusion support. At the time of discharge her condition was satisfactory, stable, afebrile with no skin rash, and no palpable lymphadenopathy. Her hemoglobin

was 9.8 g/dL, Total leucocyte count was 2280/cu mm and platelet count were 107000/cu mm.

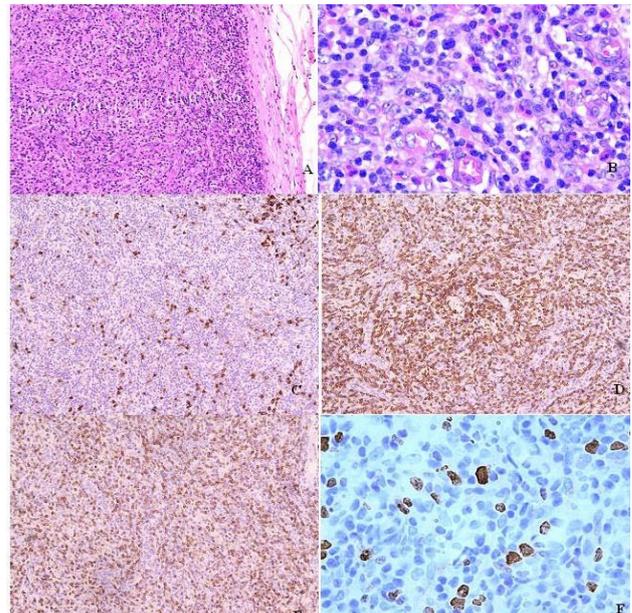


Figure 2: (a) Tissue section - lymph node biopsy showing complete effacement (H and E, X10), (b) high endothelial venules and polymorphic infiltrate - lymphocytes, immunoblasts and eosinophils (H and E, X40), (c) immunohistochemistry CD20 (X20), (d) CD3 (X20), (e) CD5 (x20), and (f) Ki-67 (x40).

DISCUSSION

The cutaneous manifestations in AITL always pose a diagnostic dilemma particularly when precede the lymph node biopsy. Their presentations are variable and described as erythematous macules and papules resembling viral exanthema or a drug reaction as seen in our case or as papulonodular eruptions, generalized petechiae, erythroderma, plaque and even as purpura fulminans.^{4,5}

Histological findings of these skin biopsies are subtle. Martel et al in his study found four types of histological pictures; mild nonspecific perivascular infiltrate of eosinophils and lymphocytes without atypia, sparse superficial perivascular infiltrate of atypical lymphocytes, dense pleomorphic infiltrate of atypical lymphocytes in superficial and deep dermis, and vasculitis without atypia. They concluded that cutaneous involvement is often related to clonal T-cell proliferation even in cases with non-specific histological pictures. But their observation was later on contradicted. Botros et al they found that identification and quantification of follicular T helper cells (Tfh) by immunohistochemistry can separate cutaneous AITL from inflammatory lesions. Again, this only suggests but cannot confirm the diagnosis of AITL. In nutshell a nodal evolution is mandatory for the diagnosis of AITL.⁶

In our case skin biopsy showed moderate perivascular, periadnexal as well as perineural and intraneural lymphohistiocytic infiltrate in both superficial and deep dermis. Some of the histiocytes are of epithelioid morphology while others are foamy along with karyorrhectic debris. Although both ZN and Wade-Fite's acid fast stains were negative on tissue sections, infective pathology could not be ruled out. These non-specific findings delayed the treatment of the patient and final diagnosis awaited till lymph node biopsy evaluation and immunohistochemistry.

Such type of diagnostic challenges in AITL are also reported in one of the largest series of cases of peripheral T-cell lymphoma where agreement on diagnosis among pathologists were only 81%.³

In view of variable non-specific histological findings on skin biopsy a conglomerate of clinical history, complementary findings, serological findings are essential and diagnosis can only be achieved by lymph node biopsy.⁷

CONCLUSION

Therefore, it has been suggested that AITL should be kept in differential diagnosis of any maculopapular lesion of unknown etiology with lymphadenopathy and lymph node biopsy and histopathological evaluation is essential for diagnosis and in best of our knowledge this is the first case of cutaneous manifestation of AITL with neural involvement.

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