CASE REPORT

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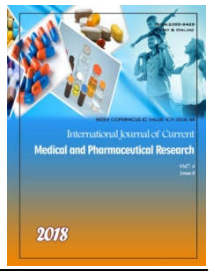

ANGIOIMMUNOBLASTIC T CELL LYMPHOMA IN A 27 YEARS OLD MAN. A RARE CASE REPORT

Deka M K * and Talukdar A

Pathology Department, Cachar Cancer Hospital & Research Centre (CCHRC), Meherpur, Silchar-788015, Assam

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ABSTRACT

2016 WHO Classification of lymphoid neoplasm has included Angioimmunoblastic T Cell Lymphoma (AITL) under the umbrella of nodal T-cell lymphoma that also include follicular T cell lymphoma and peripheral T cell lymphoma, not otherwise specified. AITL represents only 1-2% of all non hodgkin's lymphoma. A 27years old male presented with fever, pruritus and generalized lymphadenopathy. A lymph node biopsy showed the characteristic histological and immune-profile consistent with AITL. The patient received two cycles of CHOP and accomplished complete remission. A follow up period of 5 years showed recurrence free survival. The case highlights the characteristic histology and immune-profile of this rare entity of lymphoma with a particular emphasis on the efficacy of CHOP in achieving complete remission.

INTRODUCTION

2016 WHO Classification of lymphoid neoplasm has included Angioimmunoblastic T Cell Lymphoma (AITL) under the umbrella of nodal T-cell lymphoma that also include follicular T cell lymphoma and peripheral T cell lymphoma, not otherwise specified. AITL represents only 1-2% of all non hodgkin's lymphoma. AITL is an aggressive lymphoma often presenting at an advanced stage. Varied clinical features, uncharacteristic laboratory findings make the diagnosis challenging. We report a case of this rare lymphoma highlighting the characteristic histology and IHC immune-profile.

MATERIALS AND METHODS

The present study on a rare case report of Angioimmunoblastic T Cell Lymphoma in a 27 years old man was undertaken in Cachar Cancer Hospital & Research Centre (CCHRC), Meherpur, Silchar, India.

Case Report

A 27years old male from KatigorahCachar District came to the OPD with chief complaints of fever, pruritus and generalized lymphadenopathy. A FNAC done in another hospital was suggestive of Hodgkin's lymphoma. Clinical examination showed bilateral enlarged discrete firm cervical, axillary and inguinal lymph nodes. At admission abnormal laboratory findings included anemia, leukocytosis with absolute eosinophilia and elevated alkaline phosphatase level. USG abdomen showed hepato-splenomegely, multiple periportal, peri pancreatic, para aortic and mesenteric adenopathy. There was minimal ascites.

A right cervical lymph node biopsy was done. Sections showed a diffuse pattern of infiltrate completely replacing the lymph node architecture with numerous arborizing blood vessels. The infiltrate showed a polymorphous population of cells comprising of numerous atypical lymphoma cells of variable size admixed with numerous reactive cells including plasma cells, eosinophils, histiocytes, small lymphocytes and follicular dendritic cells. The blood vessels were thickened with hyperplastic endothelial cells. Occasional burned out regressive follicles were seen that showed follicular dendritic cells lacking lymphocytes. There was no presence of Reed Sternberg cells.

Immunohistochemistry showed diffuse positivity of CD3 & CD8. CD4, CD 20 & CD 30 were absent except few cells showing CD4 & CD 20 positivity. CD 21 was done that highlighted the proliferating dendritic cells.

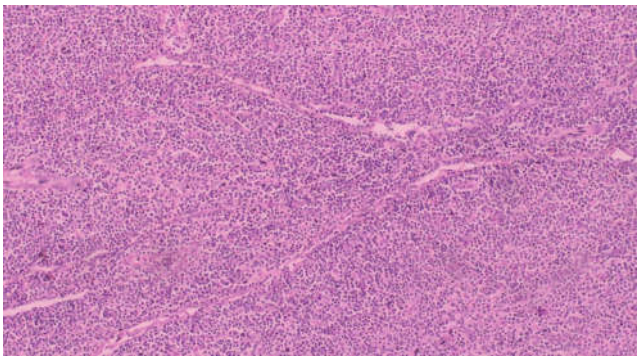
Diagnosis of angioimmunoblastic T cell lymphoma was made. Furthermore a bone marrow biopsy demonstrated lymphoma infiltration. Patient received two cycles of CHOP, tolerated well and accomplished complete remission. After follow up period of 5 years patient is alive and well.

DISCUSSION

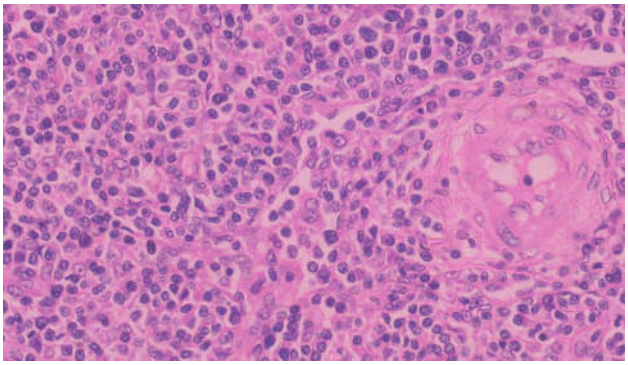
The term angioimmunoblastic lymphoma was coined in the 2008 WHO Classification of lymphoid neoplasm which was known by different names that includes angioimmunoblastic

Corresponding author:* **Deka M K
Pathology Department, Cachar Cancer Hospital & Research Centre (CCHRC), Meherpur, Silchar-788015, Assam

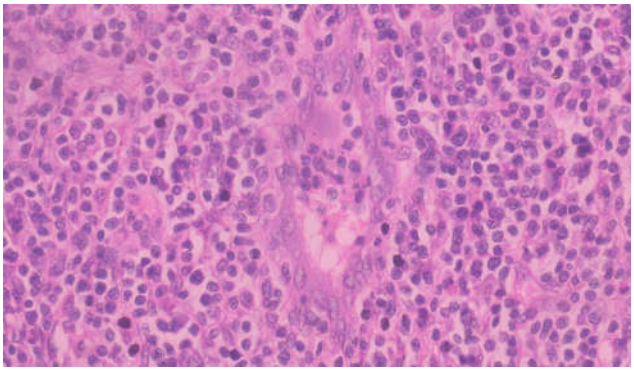
lymphadenopathy with dysproteinemia, immunoblastic lymphadenopathy and lymphogranulomatosis X. 2016 WHO Classification of lymphoid neoplasm has included AITL under the umbrella of nodal T-cell lymphoma with T follicular helper phenotype.[1,2,3,4,5]



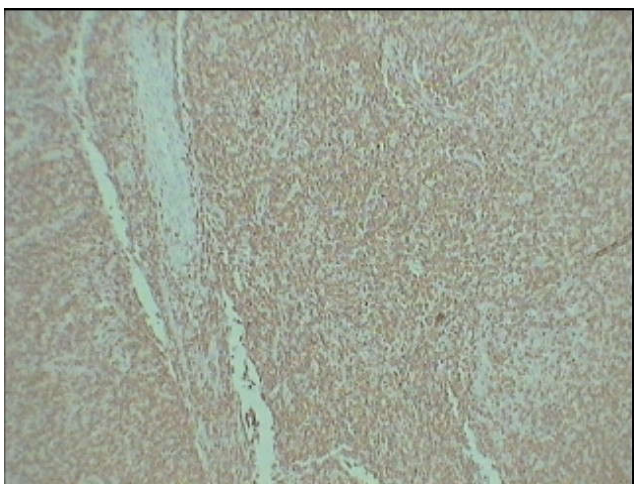
1 Photomicrographof lymph node showingdiffuse pattern of infiltrate with arborizing blood vessels.



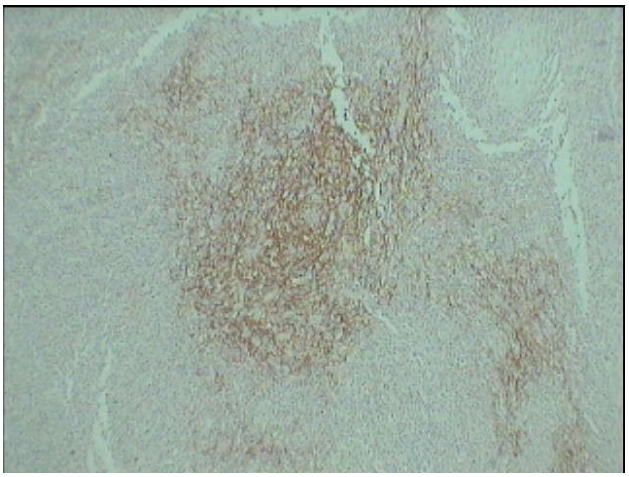
2 Photomicrograph of lymph node showing atypical lymphoid cells,admixed with numerous reactive cells and a regressive follicle.



3 Photomicrographof lymph node showing thickened vessel with hyperplastic endothelial cells.



4 Photomicrographof lymph node showing diffuse CD3 positivity.



5 Photomicrograph showing CD21 positivity highlighting the follicular dendriting cells.

AITL represents 1-2% of all non hodgskin's lymphoma and among all T cell lymphomas they constitute 18% with a slight male predominance.[6,7] There is a broad age range from 17 to 91 years with a median age of 60 years. [9,10]The most common symptoms and clinical findings at presentation includes the B-symptoms (fevers,weight loss and night sweats) and generalized lymphadenopathy. Bone Marrow involvement occurs in 70% of patients and skin manifestation in 20-50% includes rash, pruritis. Hepatoslenomegaly also noted in moderate no of patients at diagnosis.

Our patient presented with Ann Arbor clinical stage IV.
Uncharacteristic laboratory findings include elevated ESR, elevated C reactive protein, polyclonal gammopathy, leukocytosis with eosinophilia or neutrophilia, hemolytic anaemia with positive direct Coomb test, and positive rheumatoid factor.[7,8] Our patient presented with normocytic normochromic anaemia with leukocytosis and absolute eosinophilia.

Histologically there are three patterns of effacement of the lymph node architecture. While in pattern I normal lymphoid follicles can be appreciated, they show regressive changes in pattern II and III leading to 'burned out' germinal centre containing only follicular dendritic cells without lymphocytes.
Characteristic features of AILT are arborizing blood vessels which represents high endothelial venules and follicular dendritic cell expansion outside of germinal centres. A polymorphous cell population with variable number of malignant lymphoid cells in a background containing a full house of reactive cells including plasma cells, eosinophils, histiocytes, small lymphocytes and follicular dendritic cells. The neoplastic lymphoid cells have a distinctive appearance that consists of clear cytoplasm forming nodules and accumulating around blood vessels. Mitotic figures are variable. Necrosis and fibrosis are invariably absent.[7,8,11]
Our case represents the pattern III and showed the whole range of characteristic histologic findings.

AILT resembles Hodgkin's lymphoma by the fact that the malignant lymphoid cells often represents a minority component in a lymph node biopsy. AITL originate from the follicular helper T cell found in lymphoid follicle which is essential for B cell activation and differentiation. The neoplastic cell is CD 3, CD4, and CD10 positive. Characteristically they are CD4+ve and CD8-ve. In approximately 5% of cases CD4 is absent and rarely CD8

expression may be present. A more specific marker is CXCL13. Neoplastic cells are negative for CD30, CD15 or Alk-1.CD21, 23 & 35 are helpful in substantiating the important morphologic finding of follicular dendritic cell expansion.[7,8,11,12]

Our case showed the expression of pan T cell marker CD3 and T cell associated antigen CD8.CD 21 expression highlighted the follicular dendritic cell proliferation outside germinal centre. CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) chemotherapy continues to be the first line treatment of AITL. A complete remission rate of 53% is attributed to CHOP in treating AITL[8]. 5-year overall survival rate in AILT is 32% [12]Our case emphasizes the efficacy of CHOP and the patient is symptom free for last 5 years after 2 cycles of CHOP.

CONCLUSION

The diagnosis of AITL remains a challenging task. The case highlights the characteristic histology and immunoprofile of this rare entity of lymphoma with a particular emphasis on the efficacy of CHOP in achieving complete remission.

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