

# Angioimmunoblastic T cell Lymphoma: A diagnostic dilemma

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## INTRODUCTION

Angioimmunoblastic T cell lymphoma (AITL) is one of the rarest T cell lymphomas. Diagnosing the type of lymphoma is crucial, as the management differs depending on the type. We present a case of a sixty-one-year-old woman who presented with generalized lymphadenopathy and how we unfolded the diagnosis.

## CASE DESCRIPTION

Sixty-one-year old female presented with septic shock from an unknown source of infection. Ten days before the presentation she underwent excisional lymph node biopsy of left neck lymph nodes and left tonsil for generalized lymphadenopathy which she developed one month ago.

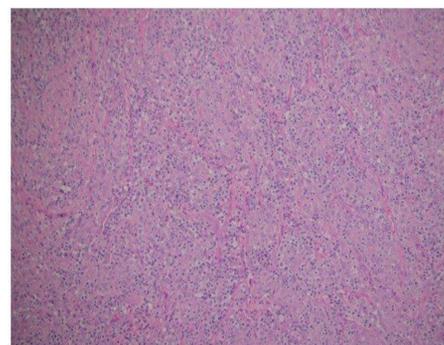
Her past medical history is significant for antiphospholipid syndrome (APLS) on warfarin. The initial workup did not reveal a source of infection. Treatment was started for septic shock with intravenous antibiotics and vasopressors.

Her microscopic sections of the biopsy showed diffusely scattered atypical large lymphocytes suspicious for large cell lymphoma. Flow cytometry did not detect any B or T-cell abnormalities.

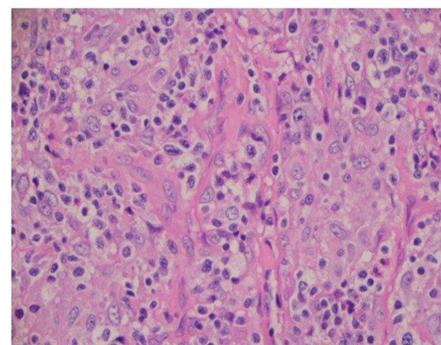
Immunohistochemistry (IHC) showed a predominance of large cells which are CD20 and CD79a positive favoring a T-cell rich B-cell Lymphoma. Gene rearrangement Clonality Analysis detected Clonal peaks 187, 193 bp which corresponds to positive for a monoclonal T- cell receptor gamma rearrangement. Also Immunoglobulin Heavy Chain (IGH) Gene Rearrangement Clonality Analysis came back negative.

Initially, we thought its a B cell lymphoma but gene rearrangement clonality analysis revealed it as T cell lymphoma and the prominent vascularity and angiocentricity favor Angioimmunoblastic T-cell Lymphoma.

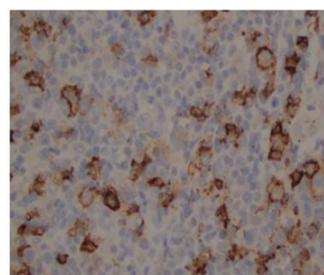
The patient remained intubated on pressor support. After the diagnosis was discussed with family, they opted for comfort care. The patient passed away within a few hours of palliative extubation.



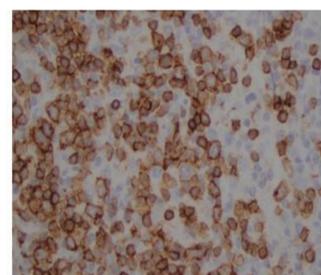
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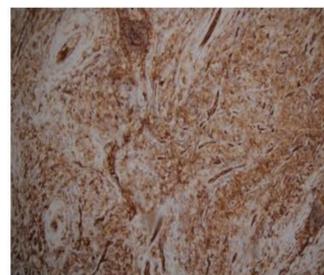
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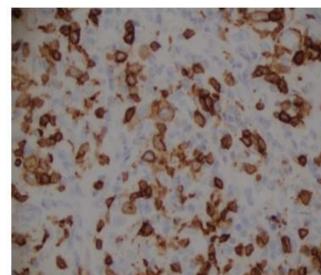
CD 3



CD 5



CD 20



CD 31

## DISCUSSION

The incidence of AITL is 0.05 cases per 100,000 person-years with a median age of 60-65 years. AITL usually presents with acute onset systemic illness associated with generalized lymphadenopathy. Type B symptoms comprise up to 85%. Diagnosis sometimes will be hard like our case and need to undergo PCR testing for gene rearrangement clonality analysis. Treatment options include steroids, combination chemotherapy, autologous hematopoietic cell transplantation (HCT), and investigational therapies. Complete remissions do occur but relapse is frequent, with median overall survival (OS) rates ranging from 15 to 36 months with chemotherapy and approximately four years with autologous HCT. Infection is the most common cause of death. Prognosis is variable with age, performance status (PS), LDH level, bone marrow involvement can independently predict survival.

## CONCLUSION

AITL are rare tumors that cause immunodeficiency due to immune dysregulation of B and T cells. Diagnosis is difficult and sometimes requires PCR for gene rearrangement clonality analysis. Treatment rarely causes remission with variable prognosis.