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Case Report

A Hiding Lymphoma: Angioimmunoblastic T Cell Lymphoma — A Case Report

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Abstract

Angioimmunoblastic T-cell lymphoma (AITL) is now a well-established subtype of mature peripheral T-cell lymphoma (PTCL). Advanced-stage disease is common with uncharacteristic laboratory and autoimmune findings that often slow or mask the diagnosis. AITL afflicts advanced-age individuals with a median age of diagnosis of 65 years of age without a notable gender predisposition. Significant strides in the immunohistochemical and molecular signature of AITL have brought increased ability to diagnose this uncommon type of PTCL. The 2016 World Health Organization classification of lymphoid neoplasms recently acknowledged the complexity of this diagnosis with the addition of other AITL-like subsets. AITL now resides under the umbrella of nodal T-cell lymphomas with follicular T helper phenotype. The treatment of relapsed or refractory AITL remains an unmet need. The spectrum of AITL from diagnosis to treatment is reviewed subsequently in a fashion that may one day lead to personalized treatment approaches in a many-faced disease. Here we report a case of angioimmunoblastic T cell lymphoma which was diagnosed using endoscopic ultrasound guided fine needle biopsy. Our patient's lab workup was unremarkable and inconclusive, but the correct diagnosis was established using endoscopic ultrasound guided fine needle biopsy. Chemotherapy started after diagnosis and his condition improved.

Keywords: *Peripheral T-Cell Lymphoma, Follicular Helper T Cells, Endoscopic Ultrasonography, Fine-Needle Biopsy.*

Introduction

Angioimmunoblastic T-Cell Lymphoma (AITL) is a peripheral T-cell lymphoma that is aggressive and has a poor prognosis. When it was originally identified as a separate entity in the 1970s, lymphoma variants were not yet recognized, and the condition was thought to be an aggressive immune response. With only 0.05 instances per 100,000 people in the US, it is an extremely rare illness [1]. It is responsible for 1-2 percent of non-Hodgkin lymphoma (HL) cases and 15-20% of PTCL cases. Fever, weight loss, urticaria, papules, red nodules, and skin lesions are the main clinical symptoms, with a median diagnostic age of less than 65 years [2].

Prodromal symptoms affect 20–50% of AITL patients, and they might show as nodular tumors or urticarial lesions on the skin. The autoimmune and unusual biochemical symptoms frequently obscure or delay diagnosis, and by the time a proper diagnosis is confirmed, the disease has frequently progressed. The pathophysiology of AITL has been revealed to be significantly influenced by Epstein-Barr virus (EBV) [2]. A median AITL survival is less than three years. To accurately diagnose this disease, excisional lymph node biopsy must be combined with concurrent immunophenotypic, molecular, and cytogenetic investigations [3] We report the case of an 80-year-old man who was diagnosed with AITL later than expected.

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Case Presentation

80 years old hypertensive male with known history of benign prostatic hyperplasia, came with a 4 weeks history of diarrhea, weight loss and fever. He denies any history of melena or hematochezia. He was non-smoker with no family history of malignancy. At the time of presentation, he was a-febrile and hemodynamically stable. The physical examination was consistent with temporal wasting, generalized abdominal tenderness and positive shifting dullness on abdominal examination. Laboratory findings were significant with normocytic anemia with hemoglobin 11.7 g/dl. His ESR was 25, CRP 109 and LDH 287. His echocardiography was normal. Imaging showed circumferential thickening and luminal narrowing of gastric antrum with perigastric fat stranding and multiple perigastric lymph nodes. Multiple enlarged perigastric nodes, upper abdominal, retroperitoneal and mesenteric lymph nodes seen on imaging.

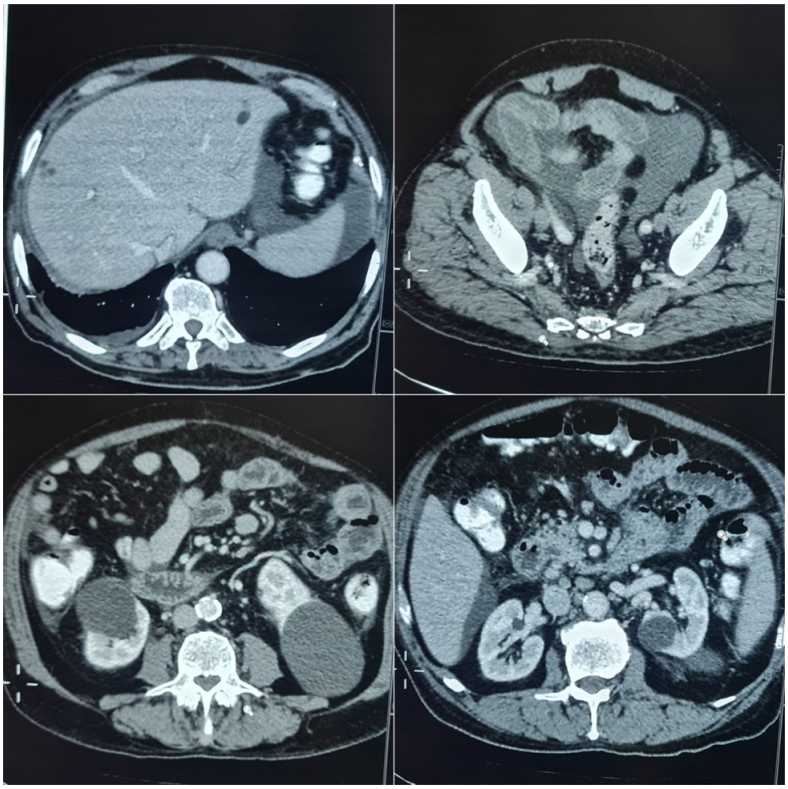

Fig 1. *Contrast Enhanced CT showing gastric thickening, ascites, multiple enlarge lymph nodes.*

There was mild abdominopelvic ascites. Findings are concerning for malignant neoplastic etiology. Colonoscopy was performed which was normal. Endoscopic ultrasound was done and 22G ACQUIRE FNB needle used and biopsy was taken from periportal lymph node.

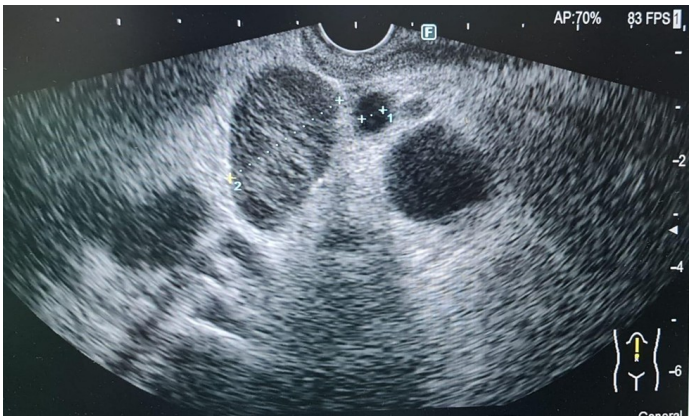

Fig 2. *Endoscopic ultrasound showing enlarged peri-portal, peri-hepatic and peri-pancreatic lymph nodes.*

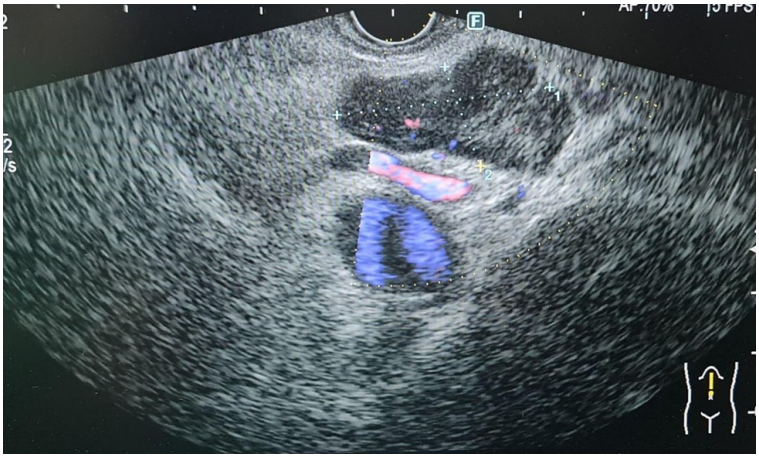

Fig 3. *Endoscopic ultrasound showing enlarge lymph nodes.*

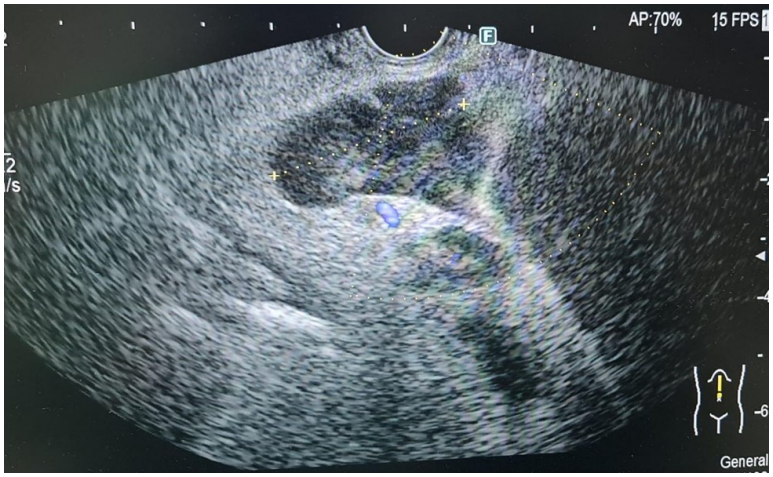

Fig 4. *Endoscopic ultrasound showing enlarged peri-hepatic lymph node.*

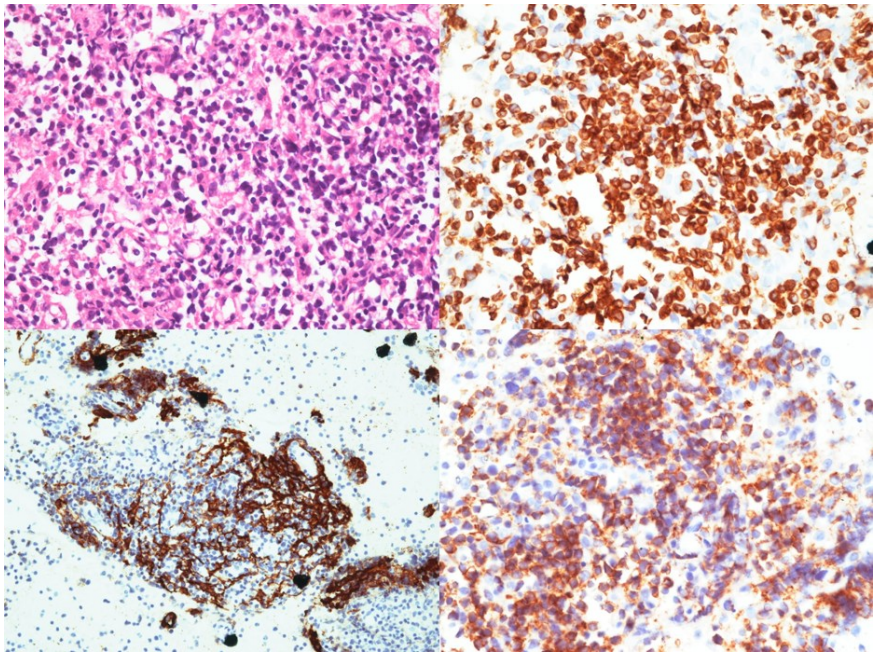

Fig 5A: *eus biopsy of lymph node showing atypical lymphoid cells.*

B: *CD3 IHC highlighting neplastic T-cell population.*

C: *CD21 highlighting follicular dendritic cell mesh work*

D: *PD1 IHC highlighting T- follicular helper cell proliferation.*

Findings were consistent with nodal T follicular helper cell lymphoma (Angioimunoblastic type). The smear showed moderately cellular and show atypical lymphoid cells with high nuclear to cytoplasmic ratio. The cell block showed fibrinous material with fragmented lymphoid tissue showing atypical lymphoid cells. The immunohistochemistry stain was positive for CD3 which was positive for atypical cells, CD30 which was patchy positive for 20% population, CD21 which highlights extra follicular distorted dendritic meshwork, CD4 which was positive for atypical cells, CD10 which was positive for atypical cells, PD1 which was positive for atypical cells, BCL6 which was patchy positive and CD20 and PAX5 which highlight B cell population. He was started on cyclophosphamide, vincristine and prednisolone. His condition and laboratory makers improved.

Discussion

Clinical, pathologic, cellular, and biologic characteristics of AITL neoplasm indicate strong inflammatory and immunological responses. Due to their phenotypic similarity to T follicular helper (Tfh) cells, tumor cells are thought to function somewhat similarly to Tfh cells, which are not cancerous and are observed in reactive follicular hyperplasia [4]. Clinical features of AITL which is a peripheral T-cell lymphoma include a high fever and widespread lymphadenopathy. The disease presentation age on average is sixty years old [5]

Similar to rheumatology, AITL has caused a diagnostic conundrum that has delayed diagnosis, treatment, and adverse prognosis. Forty to fifty percent of AITL have cutaneous involvement, with the maculopapular morbilliform rash being the most commonly encountered lesion [5]. It has a low incidence, with 0.05 new cases detected annually per 100,000 patients in the US. Europe has the highest incidence of the disease (29% of all PTL cases), followed by Asia (18%) and North America (16%); the causes of this global variability are unknown [4]. The follicular T helper cell (TFH), a subset of CD4+ T cells that controls antibody responses, is the cell of origin for AITL [6].

Due to apparent architectural preservation at lower magnification and polymorphous reactive infiltration, early stages of AITL with Attygalle pattern 1 or 2 may resemble a reactive lymph node. Making the diagnosis frequently requires a high index of suspicion in addition to experience [6]. AITL can be accompanied by biochemical abnormalities such as high C-reactive protein, elevated lactate dehydrogenase (LDH), elevated creatinine, hypergammaglobulinemia, and hypoalbuminemia. As in our instance, 16% of cases had an initial histological misinterpretation of the lymph node sample as reactive lymphadenopathy [5]. The current patient was reported at a later stage of the illness and had numerous symptoms that resembled an infectious condition, including weight loss, a prolonged fever, and lymphadenopathies [4]

Although the majority of patients arrive with multisystem involvement and an advanced stage of the disease, reports of the disease spreading to the peripheral circulation have not always been confirmed. The most popular intense regimen in PTCL is still the CHOP regimen [7]. The addition of etoposide to CHOP (CHOEP) shows an overall response rate (ORR) of 82% and a complete response (CR) of 51% in the management of PTCL and better progression-free survival (PFS) in young and fit patients when compared to CHOP alone. etoposide has a reported 53% complete response (CR) when used as a first-line therapy for AITL. Those above 60 years of age, however, are more toxic to it. Our patient was prescribed CHOEP as a first-line treatment based on the evidence that was available. Patients with relapse/refractory (R/R) disease should have their eligibility for autologous stem cell transplantation (ASCT) carefully evaluated as part of their therapy [7].

In conclusion, this example highlights the difficulties with AITL diagnosis. There are occasionally specific triggers for AITL, such EBV infection, medication use, and so forth. AITL frequently coexists with other illnesses, particularly immune -related conditions and malignancies. Furthermore, the identification of genetic changes, such as abnormal TCR rearrangements or common chromosomal aberrations by fluorescence in situ hybridization, and certain immune-histochemical testing may make the diagnosis of underlying AITL easier [8].

Conclusion

This case report illustrates the diagnostic challenge associated with AITL. AITL is an aggressive T-cell lymphoma with a poor overall prognosis. Combination chemotherapy is typically offered to these patients, and those who respond well to initial therapy are considered for autologous stem cell transplantation.

Conflict of Interest

None

Funding Disclosure

None

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