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Angioimmunoblastic T Cell Lymphoma

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Dear Editor,

Angioimmunoblastic T cell lymphoma (AITL) is one of the most common Peripheral T cell lymphomas (PTCL) arising from a subset of peripheral CD4 positive T cells but is a rare form of Non-Hodgkin lymphoma. AITL was first described as an angioimmunoblastic lymphadenopathy with dysproteinemia in 1974[1]. The incidence of AITL is higher in Europe (28% of PTCL) than in the USA (15% of PTCL) and in Asia (17% of PTCL). Patients frequently exhibit B-symptoms and generalized lymphadenopathy. Other clinical features include hepatomegaly, splenomegaly, polymorphic skin rash and pleural effusion. Advanced stage disease (Ann Arbor III/IV) is observed in 80% of cases. Polyclonal hypergammaglobulinemia occurs in approximately 50 percent of AITL cases[2].

There is no standard treatment for AITL. Corticosteroids have been the first line agents and can be given alone or in combination with cyclophosphamide, vincristine, or both. CHOP and CHOP based regimens have produced complete remission rates of 60% in retrospective analysis. In 2018, brentuximab was approved by the FDA for CD 30 expressing PTCLs. Brentuximab plus CHP was superior to CHOP for progression-free survival (p=0.011). Furthermore, the natural history of AITL is characterized by several relapses, with a five-year overall survival of 30 percent[3]. The survival is significantly related to age, stage and clinical features. Several case reports have been published in recent years showing the complexity and aggressive nature of the disease[4].

This letter discusses the complex presentation of a confirmed case of AITL in Ireland. This is a case of a 75-year old male who presented with lethargy, shortness of breath and productive cough for the past few months. He was a chronic smoker and had no significant background history. Physical examination revealed bilateral submandibular, cervical, axillary and inguinal lymphadenopathy. His breath sounds were reduced at both lung bases and, he had moderate ascites with peripheral edema.

Blood tests showed anemia, thrombocytopenia, elevated ESR with deranged renal function. Chest x-ray revealed bi-basal consolidations with small pleural effusions. CT thorax, abdomen and pelvis was performed and showed diffuse adenopathy, bilateral pleural effusions, bi-basal consolidation, moderate splenomegaly and moderate ascites. He underwent excision biopsy of left cervical lymph nodes and histopathology showed T cell lymphoma with expression of T follicular helper cell markers, regarded as angioimmunoblastic T cell lymphoma.

His bone marrow aspirate was taken, and it revealed an increased population of T lymphocytes. Other tests for relevant differentials including quinterferon test, autoimmune screen, vasculitic screen, hepatitis and HIV serology, which were all negative.

Sepsis protocol was commenced as initial management of the patient and relevant investigations were carried out to find severe underlying illness. On reaching the final diagnosis, the patient received the first cycle of chemotherapy (CHOP) along with supportive measures. With further progression of the disease to an advanced stage, the patient's condition didn't improve. A decision was made to consider palliative treatment and stop further chemotherapy as patient developed multi-organ failure, but unfortunately, he died within a short period of time.

This case illustrates the aggressive nature of AITL and explains the complexity of the disease as it can mimic infectious, autoimmune and allergic aetiologies. At the time of diagnosis, most of the patients present with advanced-stage disease (stages III-IV). The diagnosis of AITL can be challenging, given the lack of clinical and histological diagnostic criteria therefore, clinical history, symptoms and complementary studies are crucial.

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