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# Case Report

## A Case of Angioimmunoblastic T-cell Lymphoma was Diagnosed Initially as Kimura's Disease and Castleman Disease

Md Shafiul Kabir[1], Amirah Mahboob[1], Md Rajilur Rashed[1], Tahsina Tasneem Isnat[1], Subrata Kumar Biswas[1], Naima Afroza[1], Md. Atikul Islam[2], Md. Rashedul Hasan[2], Md. Maidul Islam[1], Mohammad Mahfuzul Hoque[2], Md. Shafiqul Bari[2]

**Abstract:**

*Kimura disease (KD) is a rare chronic inflammatory disorder, and Castleman disease is a rare lymphoproliferative syndrome. Castleman disease (CD) is associated with an increased risk for lymphoma. The patient described here is a 52-year-old female who came with one-year history of fever, generalized lymphadenopathy and polyserositis. The patient was initially diagnosed on 2 occasions respectively as Kimura's disease and then as Castleman disease. Eventually, she had a confirmed diagnosis of Angioimmunoblastic T-cell lymphoma, after which she received 2 cycles of combination chemotherapy and her condition improved. She received a total of 6 cycles of chemotherapy over the next 6 months, but unfortunately, she passed away after receiving the 6th cycle.*

***Keywords:*** *Kimura's Disease, Castleman Disease, Angioimmunoblastic T-cell lymphoma, Lymphadenopathy*

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**Introduction:**

Kimura disease (KD) is a rare chronic inflammatory disorder of unknown etiology. It usually presents as subcutaneous mass in the head and neck region and is frequently associated with regional lymphadenopathy or salivary gland involvement.[1,2] The commonly involved sites are periauricular, groin, orbit, and eyelids.[3] Castleman disease, first described in 1956, is a lymphoproliferative syndrome known as angiofollicular lymph node hyperplasia. Its incidence is estimated at 25 cases per million persons annually.[4]. Castleman disease can progress to, or it may be associated de novo with, malignant lymphoma, which makes both the diagnosis and treatment complex.[5] The association between Lymphoma and Castleman's disease is rare and well-documented. The cases reported in the literature of Hodgkin's lymphoma frequently were of the interfollicular subtype and coexisted with the multicentric plasma cell variant of Castleman's disease.[6] Angioimmunoblastic T-cell lymphoma (AITL) is an aggressive peripheral T-cell lymphoma with a poor prognosis.[7] According to the International Prognostic Index (IPI), most patients present with intermediate or high-risk disease, and high-risk patients have a five-year survival rate of approximately 20%.[8] There is no standard of care for the treatment of AITL, although lower-risk patients may be offered treatment with high-dose steroids. In contrast, patients with higher-risk diseases are often treated with an anthracycline-based combination regimen such as CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) .[9] Patients who achieve a complete response (CR) with initial therapy are often considered for autologous hematopoietic stem cell transplant (HSCT).[10,11]

We report here a case which was initially diagnosed as Kimura's disease, then later as multicentric Castleman disorder on histopathology, but ultimately, as the clinical feature suggested, was diagnosed as Angioimmunoblastic T-cell Lymphoma on immunohistochemistry.

**Case Report:**

A 52-years-old, normotensive, non-diabetic, married Muslim woman hailing from Cumilla got admitted into DMCH with the complaints of fever, productive cough, epigastric pain, breathlessness and heaviness over chest for 8 months and ulceration of oral mucosa and tongue for 15 days and had associated anorexia and weight loss. Fever was initially low-grade, intermittent with evening rise of temperature for the first 2 months, but later on became high-grade, continued but was not associated with night sweats or chills & rigor. She

1. Department of Medicine, Dhaka Medical College Hospital, Dhaka, Bangladesh
2. Department of Medicine, Dhaka Medical College, Dhaka, Bangladesh

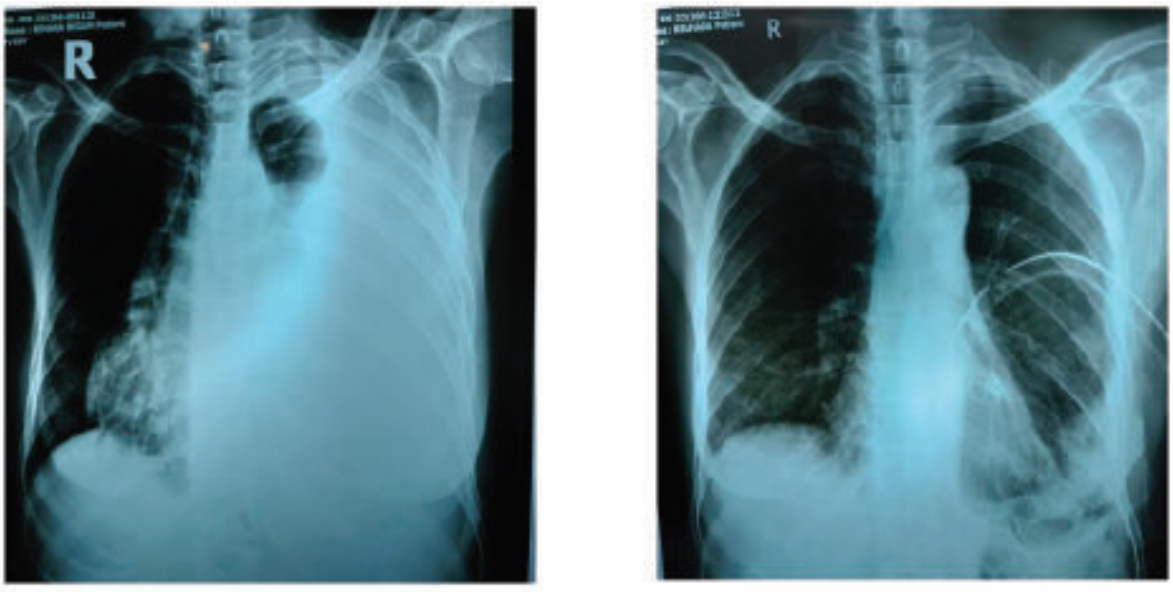
**Corresponding author:** Dr. Md Shafiul Kabir, Department of Medicine, Dhaka Medical College Hospital, Dhaka, Bangladesh. Email: sagor9364@gmail.com

gave no history of contact with TB patient, hemoptysis, night sweat, joint pain, rash, dysphagia, vomiting, hematemesis, melaena, itching, jaundice, epistaxis, gum bleeding and altered bowel habit. On examination, the patient was anemic and had angular stomatitis with ulceration of oral mucosa and tongue. There was bilateral pitting oedema and generalized lymphadenopathy involving posterior cervical and inguinal region of both sides. Lymph nodes were multiple, discrete, firm, non-tender of variable size and shape, largest one being in right inguinal region which is 5x 3 cm. They were not fixed with underlying structure or overlying skin and had no discharging sinus. She had an intercostal tube in situ over left chest and there was evidence of left sided pleural effusion (Figure 1). There was an epigastric lump palpable per abdomen of 3x2 cm, having smooth surface, tender, firm in consistency and fixed with underlying structure but not overlying skin. Ascites was present as evidenced by shifting dullness. Over the course of her illness, the patient had been admitted into local hospital multiple times and received multiple courses of antibiotics including anti-TB drugs empirically for 3 weeks with no response. She also gave history of thoracentesis 13 times during this time from left chest for relief of symptoms. Subsequently 5 months back she was admitted to Cumilla Medical College. There, she was diagnosed with a case of Kimura's disease as per histopathology and immunohistochemistry from the left supraclavicular lymph node and was referred to DMCH for further management. She was admitted to the Department of Hematology on 3rd December 2022 and was treated with immunosuppressants as per the treatment protocol for Kimura's disease, but she did not respond to the treatment. She was then transferred to the Department of Medicine on 14th December 2022. Previously, the patient had anaemia only. Subsequently, the patient developed cytopenia in the form of anaemia and leukopenia. The patient also had hyponatremia and hypoalbuminemia. A pleural fluid study was done, which was revealed to be transudative in nature, but no conclusive findings were found. A pleural biopsy was done, and only chronic inflammatory cells were found, indicating chronic lymphadenitis. CT scan of the abdomen revealed abdominal lymphadenopathy. CT-guided FNAC from the abdominal lymph node was done, the report of which was inconclusive. Serum IgG4 level was also done which was expected. After facing such a diagnostic dilemma, a repeat lymph node biopsy was done from the inguinal lymph node. Surprisingly, the histopathology revealed Castleman's disease, a hyaline vascular type. However, the immunohistochemistry of the tissue was tested for 16 markers, and the findings were consistent with Angioimmunoblastic T-cell Lymphoma.

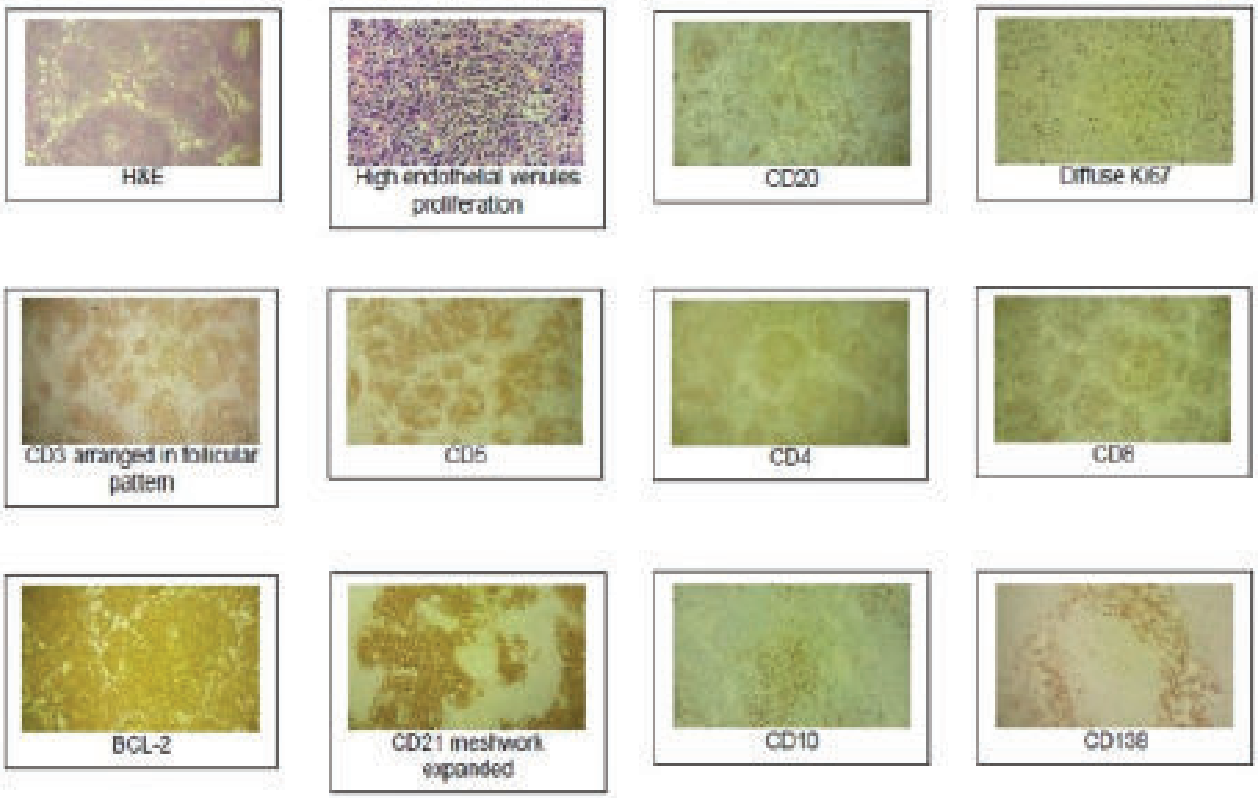
According to the treatment protocol of angioimmunoblastic T-cell Lymphoma, the patient received 2 cycles of chemotherapy consisting of CHOP regimen+Etoposide therapy and the patient's fever and pleural effusion subsided after 2 cycles of chemotherapy. However, after six cycles of chemotherapy completed, the patient sadly passed away on July 16th, 2023.

**Table 1.** Complete Blood Count showing anemia progressing to bicytopenia

| | 4-12-22 | 14-12-22 | 23-12-22 | 12/01/23 |
|---|---|---|---|---|
| Hb (gm/dl) | 7.8 | 8.9 | 4.9 | 9.4 |
| ESR(mm in 1st hr) | 20 | 24 | 18 | 16 |
| WBC | 6,500 | 2,250 | 2,100 | 3,650 |
| Platelet | 2,42,000 | 46,000 | 1,60,000 | 1,90,000 |
| Neutrophil (%) | 57 | 57.7 | 57 | 73 |
| Lymphocyte (%) | 39 | 34.7 | 32 | 20 |
| MCV (fl/L) | 81.7 | 78.5 | 86.9 | 84 |




Chest X-ray P/A view showing pleural effusion, before intercostal tube insertion (left), after intercostal tube insertion (right)




**HISTOPATHOLOGY FINAL DIAGNOSIS PANEL**
(Immunohistochemistry)

| IHC MARKER(S) | RESULT |
|---|---|
| CD20 | Focally positive in B cells (3+) |
| CD30 | Occasional immunoblasts are positive (1+) |
| CD21 | Few dendritic cell meshwork are broken and in another lymph node follicular dendritic cell meshwork expanded (4+) |
| Tdt | Negative (0%) |
| LMO2 | Negative (0%) |
| BCL-2 | Strong and diffuse positive (4+) |
| CD3 | Positive in follicular / nodular pattern (4+) |

| | |
|---|---|
| BCL-6 | Focally positive (1+) |
| Ki67 | Diffuse positive (2+) |
| CD5 | Positive in follicular / nodular pattern (4+) |
| CD4 | Positive in follicular / nodular pattern (4+) |
| CD8a | Positive in follicular / nodular pattern (4+) |
| PD1 | Negative (0%) |
| CD138 | Positive in follicular plasma cells (1+) |
| HHV-8 | Negative (0%) |
| CD10 | Positive (3+) |

**Figure 2.** Findings of nodular aggregates of T-lymphocytes with high endothelial venule proliferation with expansion of CD21 positive FDC meshwork, diffuse Ki67, BCL-2, CD10, CD138 positivity indicates a diagnosis in favor of Angioimmunoblastic T-cell lymphoma

**Discussion:**

This case report describes a rare association between benign lymphoproliferative disease, such as Kimura's disease, and multicentric Castleman's disease with malignant lymphoproliferative disease, such as Lymphoma. Despite the histopathology of lymph nodes revealing Kimura's disease and Castleman's disease on two separate occasions, the patient was not responsive to immunosuppressive treatment & the patient's clinical scenario was more consistent with Lymphoma. This drove us to explore further, revealing the underlying aggressive Angioimmunoblastic T-cell Lymphoma. The prognosis of this disease is inferior, so early diagnosis & treatment is a must.

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