Fever With Lymphadenopathy: A Rare Case Of Kikuchi-Fujimoto Disease

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Abstract: **Kikuchi-Fujimoto Disease (KFD), also known as histiocytic necrotizing lymphadenitis, is a rare, benign, and self-limiting condition typically affecting young women. Its clinical presentation often mimics other infectious, autoimmune, or malignant causes of lymphadenopathy such as tuberculosis, systemic lupus erythematosus (SLE), or lymphoma, thereby complicating timely diagnosis. We report a unique case of KFD in a 32-year-old female who presented with fever, lymphadenopathy, and autoimmune features consistent with an overlap syndrome involving SLE and Scleroderma, suggestive of Mixed Connective Tissue Disease (MCTD). Histopathological examination of lymph nodes revealed characteristic features of KFD. The case highlights the need for a high index of clinical suspicion, thorough differential diagnosis, and the critical role of lymph node biopsy to avoid misdiagnosis and unnecessary treatments.**

I. INTRODUCTION

Kikuchi-Fujimoto Disease (KFD) was first described in Japan in 1972 and is characterized histologically by necrotizing lymphadenitis predominantly involving histiocytes and CD8+ T cells without granulocytic infiltration. It is commonly seen in Asian women under the age of 40 and has an indolent, self-limiting course lasting several weeks to months. While the exact etiology is uncertain, proposed mechanisms include viral triggers (e.g., Epstein-Barr Virus, Human Herpesvirus) and autoimmune predispositions such as SLE or other connective tissue disorders.

The diagnostic challenge lies in differentiating KFD from conditions with similar presentations, especially in TB-endemic areas. Importantly, KFD has been reported in association with autoimmune diseases, most commonly SLE, either concurrently or sequentially. Here, we report a rare case of KFD in a patient with features suggestive of MCTD involving SLE and Scleroderma.

II. CASE REPORT

A 32-year-old woman recently returned from Dubai presented with:

- 15 days history of high-grade fever associated with Night sweats
- Productive cough with yellowish sputum since 1 week
- Painful right-sided cervical and axillary swelling since 1 week

No similar complaints in the past , No comorbidities , no addictions . no significant family history
On general examination, pt is pallor with lymphadenopathy
Other features

- Malar rash
- Sclerodactyly
- Beaked nose
- Puckered mouth
- Right cervical and axillary lymphadenopathy

On systemic examination, CVS , CNS . Abdominal and Respiration system were normal

III. INVESTIGATIONS

Laboratory:

- Hemoglobin: 9.1 g/dL
- Total Leukocyte Count: 5,700/µL (Neutrophil predominance)
- ESR: 120 mm/hr (markedly elevated)
- ANA: ≥1:1280
- Anti-dsDNA: Positive
- Anti-Smith: Positive
- U1-RNP: Positive
- Ribosomal P: 184.0 U/mL
- EBV serology: Negative
- Sputum ZN stain and CBNAAT: Negative
- Serum creatinine: 1.0 mg/dL
- Urine analysis: Normal

Imaging:

- Chest X-ray: Normal
- USG Neck: Multiple enlarged cervical lymph nodes, up to 2.8 x 1.1 cm, preserved fatty hilum
- USG Abdomen: Mild hepatomegaly

Histopathology: Biopsy of the right axillary lymph node revealed:

- Necrotizing lymphadenitis with histiocytes, lymphocytes, and patchy necrosis
- No granulomas or malignant cells
- Findings consistent with Kikuchi-Fujimoto Disease

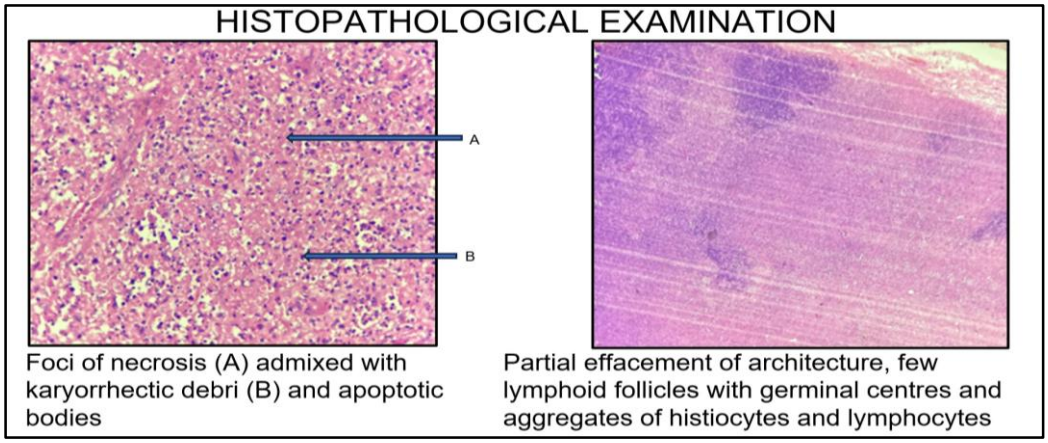

Fig: histopathological slide of axillary lymph node biopsy

IV. RESULTS

Based on clinical presentation and investigative findings, the primary diagnosis was Kikuchi-Fujimoto Disease (KFD), confirmed through histopathological examination of the axillary lymph node showing necrotizing lymphadenitis without granulomas or malignancy. Concurrently, the autoimmune profile revealed elevated ANA titers with anti-dsDNA, Smith, and U1-RNP positivity, consistent with an overlap syndrome—Mixed Connective Tissue Disease (MCTD) involving Systemic Lupus Erythematosus (SLE) and Scleroderma.

V. MANAGEMENT AND OUTCOME

The patient was initiated on systemic corticosteroids along with supportive therapy. Her symptoms, including fever, lymphadenopathy, and constitutional complaints, resolved entirely within six weeks. This favorable response highlights the self-limiting nature of KFD and suggests that early corticosteroid intervention may be particularly beneficial in cases associated with autoimmune overlap syndromes, aiding in rapid clinical recovery.

VI. DISCUSSION

Kikuchi-Fujimoto Disease (KFD) presents a significant diagnostic challenge due to its clinical similarities with infectious diseases, such as tuberculosis, and malignancies like lymphoma. This is especially problematic in tuberculosis-endemic regions, such as India, where persistent lymphadenopathy and systemic symptoms often lead to misdiagnosis and unnecessary anti-tuberculosis treatment. This case highlights the need to consider KFD in the differential diagnosis of patients with persistent lymphadenopathy and systemic symptoms. KFD is frequently associated with autoimmune disorders, particularly Systemic Lupus Erythematosus (SLE). Studies have shown that KFD can occur before, during, or after the diagnosis of SLE, suggesting a shared autoimmune mechanism. In our patient, the presence of positive autoantibodies (ANA, anti-dsDNA, anti-Smith, and U1-RNP) and clinical features of both SLE and Scleroderma led to the diagnosis of Mixed Connective Tissue Disease (MCTD), reinforcing this connection.

Histopathology remains the gold standard for diagnosis, with characteristic findings including necrotizing lymphadenitis and apoptotic debris. While KFD is usually self-limiting, corticosteroid therapy can aid in recovery when associated with autoimmune conditions, as demonstrated in this case.

VII. CONCLUSION

Kikuchi-Fujimoto Disease should be included in the differential diagnosis of fever and lymphadenopathy, particularly in young women with autoimmune

features. Histopathology is crucial to confirm diagnosis and prevent unnecessary treatment. The association with SLE and MCTD underscores the need for vigilant follow-up. With appropriate management, the prognosis remains excellent.

VIII. CONSENT

The consent was taken and signed by respective patient.

IX. CONFLICTS OF INTEREST

The authors have no conflicts of interest.

X. REFERENCES

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