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Kikuchi-Fujimoto Disease Presenting as Acute Febrile Pharyngo-Tonsillitis in a Young Female: A Rare Clinical Masquerade

Dr. Harikrishna Sharma[1], Dr. Prashant Patil[2], Dr. Swapnil Gautam[3]

[1]DNB Junior Resident, Department of General Medicine, Bhaktivedanta Hospital and Research Institute, Mira Road Thane, Maharashtra. 401107
Corresponding Author Email: *harikrishnahari4[at]gmail.com*

[2]Consultant Physician, Department of General Medicine, Bhaktivedanta Hospital and Research Institute, Mira Road Thane, Maharashtra. 401107

[3]Consultant Physician & Infectious Disease Specialist, Department of General Medicine, Bhaktivedanta Hospital and Research Institute, Mira Road Thane, Maharashtra. 401107

Abstract: ***Kikuchi–Fujimoto disease (KFD) is a rare, benign, self-limiting cause of cervical lymphadenopathy and fever, predominantly affecting young women[1]. Due to its nonspecific clinical presentation, it is frequently misdiagnosed as tuberculosis, lymphoma, or systemic lupus erythematosus, especially in tuberculosis-endemic regions. Presentation as acute febrile pharyngo-tonsillitis is uncommon and may lead to diagnostic delay and inappropriate antimicrobial therapy. We report the case of a 24-year-old female who presented with prolonged fever and acute pharyngo-tonsillitis, initially treated as bacterial infection. Persistence of fever and cervical lymphadenopathy despite antibiotic therapy prompted lymph node biopsy, which confirmed Kikuchi–Fujimoto disease[4]. Early recognition of this atypical presentation is essential to avoid unnecessary investigations and prolonged antimicrobial use.***

Keywords: Kikuchi–Fujimoto disease; necrotizing lymphadenitis; pharyngo-tonsillitis; cervical lymphadenopathy; fever

1. Introduction

Kikuchi–Fujimoto disease is a rare inflammatory disorder characterized by histiocytic necrotizing lymphadenitis[1]. It typically presents with fever and tender cervical lymphadenopathy and predominantly affects young women. Owing to its overlapping clinical features with tuberculosis, lymphoma, and autoimmune disorders, Kikuchi disease is often misdiagnosed, particularly in regions where tuberculosis is endemic, such as India[3]. Although cervical lymphadenopathy is a hallmark feature, initial presentation with ENT manifestations such as pharyngitis or tonsillitis is

rare and poorly recognized[5]. We report an unusual case of Kikuchi–Fujimoto disease presenting as acute febrile pharyngo-tonsillitis, leading to initial diagnostic confusion.

2. Case Report

A 24-year-old previously healthy female presented with high-grade intermittent fever for 12 days, associated with severe sore throat, odynophagia, and bilateral calf pain. There was no history of cough, weight loss, night sweats, skin rash, photosensitivity, joint pain, oral ulcers, or prior tuberculosis exposure.

On admission, the patient was febrile and hemodynamically stable. Oropharyngeal examination revealed a congested pharynx with inflamed tonsils[3.] without exudates. Neck examination showed multiple tender, firm, mobile lymph nodes in the left cervical region (levels II and III). There was no hepatosplenomegaly, skin rash, or arthritis. The remainder of the systemic examination was unremarkable.

Initial laboratory investigations revealed hemoglobin of 11.7 g/dL, total leukocyte count of 11,260/mm³, ESR of 67 mm/hr, and C-reactive protein of 72 mg/L. Liver and renal function tests were within normal limits. Two sets of blood cultures were sterile.

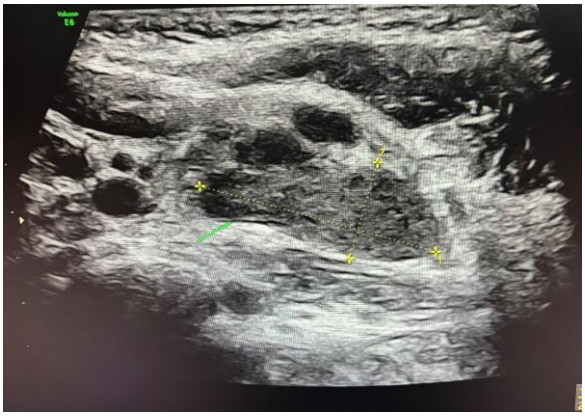
The patient had received multiple courses of oral antibiotics prior to admission for presumed acute bacterial pharyngo-tonsillitis[3.], without clinical improvement. She was initiated on intravenous piperacillin–tazobactam (4.5 g every 8 hours), along with supportive care including intravenous fluids, betadine gargles, and NSAIDs. Although symptoms of pharyngo-tonsillitis improved within two days, high-grade fever and cervical lymph node tenderness persisted.

Ultrasonography of the neck demonstrated multiple enlarged hypoechoic cervical lymph nodes with loss of fatty hilum, the largest measuring 2.5 × 1.0 cm. Further evaluation revealed mildly elevated serum ferritin levels. Antinuclear antibody was positive at a titer of 1:100 with a cytoplasmic pattern, while anti-double-stranded DNA, anti-CCP antibodies, and rheumatoid factor were negative. Viral serology for HIV, Epstein–Barr virus, and cytomegalovirus was negative.

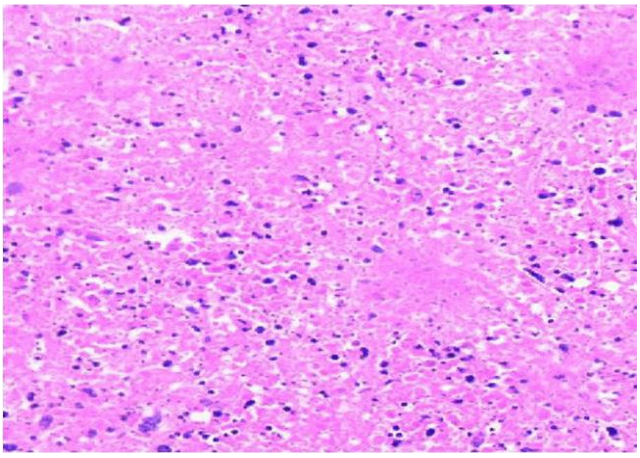
Given persistent fever beyond five days of hospitalization, an ultrasound-guided left cervical lymph node biopsy was performed. Histopathological examination revealed areas of necrosis with abundant karyorrhectic debris, numerous histiocytoid cells, and absence of neutrophils or granulomas. Viable lymphoid cells and dendritic cells were also noted, with inflammation extending into the perinodal adipose

tissue. These findings were consistent with necrotizing lymphadenitis suggestive of Kikuchi- Fujimoto disease[4]. GeneXpert testing, acid-fast bacilli smear, bacterial cultures, and TB-MGIT cultures were negative.

Antibiotics were discontinued, and the patient was treated with a single dose of intravenous dexamethasone 8 mg along with oral NSAIDs[5]. She showed marked clinical improvement within 48 hours, with complete resolution of fever and neck pain over the next three days. She was discharged on oral NSAIDs. On follow-up after 10 days, cervical lymphadenopathy had resolved completely, and the patient remained asymptomatic[6].



USG -Neck showing Multiple Hypoechoiec
Lymph nodes Largest measuring 2.5*1cm (Green arrow)



Lymph Node biopsy showed areas of necrosis with karyorrhectic debris, numerous histiocytoid cells, & absence of neutrophils or granulomas, consistent with necrotizing lymphadenitis.

3. Discussion

Kikuchi–Fujimoto disease[6.] is an uncommon cause of prolonged fever and cervical lymphadenopathy in young adults. The disease is frequently misdiagnosed as tuberculosis, lymphoma, or systemic lupus erythematosus due to overlapping clinical and laboratory features. ENT manifestations are rare, and presentation as acute febrile pharyngo-tonsillitis can easily mislead clinicians toward a diagnosis of bacterial infection, as observed in the present case.

Failure to respond to appropriate antibiotic therapy, persistent fever, and progressive lymphadenopathy should prompt evaluation for alternative diagnoses. Histopathological examination of lymph node tissue remains the gold standard for diagnosis.

Transient antinuclear antibody positivity has been reported in a subset of patients with Kikuchi disease. In the absence of clinical and immunological criteria for systemic lupus erythematosus, ANA positivity is considered incidental, though long-term follow-up is recommended.

Kikuchi- Fujimoto disease is usually self-limiting[7.], and most patients respond well to symptomatic treatment with NSAIDs. Corticosteroids are reserved for patients with severe symptoms, persistent fever, or extranodal involvement. Early recognition of this condition helps avoid unnecessary antimicrobial therapy, invasive investigations, and patient anxiety.

4. Conclusion

Kikuchi- Fujimoto disease can present atypically as acute febrile pharyngo-tonsillitis[8.], leading to diagnostic delay and inappropriate treatment. Persistent fever associated with cervical lymphadenopathy, particularly in young women, should prompt early consideration of Kikuchi disease and timely lymph node biopsy. Awareness of this rare presentation is essential for accurate diagnosis and optimal patient management.

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