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A Wolf in Sheep's Clothing: Kikuchi-Fujimoto Disease Masquerading as Tubercular Lymphadenopathy: A Case Report

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Abstract

Kikuchi-Fujimoto Syndrome (KFS), also known as histiocytic necrotizing lymphadenitis, is a rare, self-limiting disorder primarily affecting young adults, with a higher prevalence in Asian populations. It presents with fever, lymphadenopathy, and leukopenia, often mimicking infectious (Tuberculosis), autoimmune, or malignant diseases. The exact etiology remains unclear, though viral and autoimmune mechanisms are proposed. Diagnosis is confirmed through histopathological examination of lymph node biopsy, which reveals characteristic necrosis and histiocytic infiltration without neutrophils. Treatment is usually supportive, with nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids used for symptomatic relief. This paper presents a case of a 28-year-old female who was initially misdiagnosed with tubercular lymphadenopathy and subsequently diagnosed with KFD. and a comprehensive review of the current literature on KFS, emphasizing diagnostic challenges and management strategies.

Keywords: Kikuchi-Fujimoto Syndrome, Histiocytic Necrotizing Lymphadenitis, Lymphadenopathy, Autoimmune Disorder, Differential Diagnosis

Categories: Internal Medicine, Infectious Disease

Introduction

Kikuchi-Fujimoto Syndrome (KFS) was first described in Japan in 1972. It is a rare, benign, and self-limiting disorder characterized by fever and lymphadenopathy[1]. Due to its clinical and histopathological similarities to other serious conditions, including tuberculosis, lymphoma, and systemic lupus erythematosus (SLE), KFS presents a significant diagnostic challenge [2,3]. This study aims to present a case report and review recent

advancements in the understanding and management of this rare entity.

Case Presentation

A 28-year-old female presented with complaints of fatigue, fever, night sweats, and a mild cough with minimal expectoration for one month. On physical examination, cervical lymphadenopathy was noted. Fine- needle aspiration cytology (FNAC) of the affected lymph nodes revealed granulomatous inflammation, leading to a preliminary diagnosis of tubercular lymphadenopathy. Consequently, the patient was initiated on Category I anti-tubercular therapy (ATT).

After one week of treatment, the patient reported symptomatic relief. However, lymphadenopathy persisted beyond two weeks, prompting an excisional biopsy of the lymph node. Histopathological analysis revealed necrotizing lymphadenitis, consistent with Kikuchi-Fujimoto disease. ATT was discontinued following confirmation of the diagnosis. The patient was managed with nonsteroidal anti-inflammatory drugs (NSAIDs) and a short course of corticosteroids, resulting in complete clinical resolution.

During a follow-up evaluation at two months, an antinuclear antibody (ANA) profile was performed to assess a potential association with systemic lupus erythematosus (SLE). The ANA test was negative, ruling out any concurrent autoimmune pathology. The patient remained asymptomatic and in good health at subsequent follow-up.

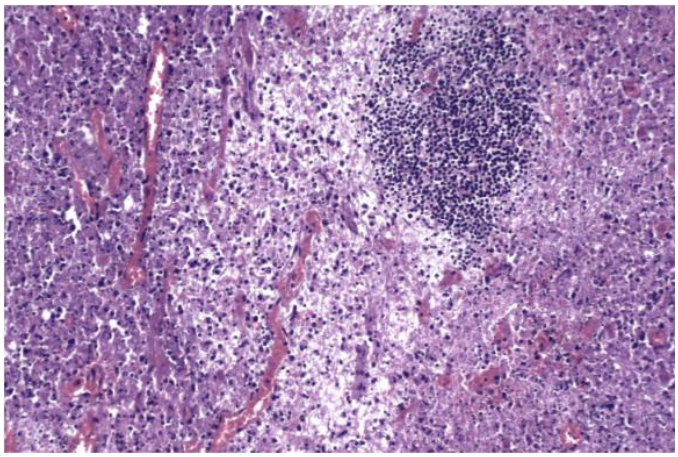

Figure 1: Pale areas composed of histiocytes, eosinophilic granular material, and nuclear debris.

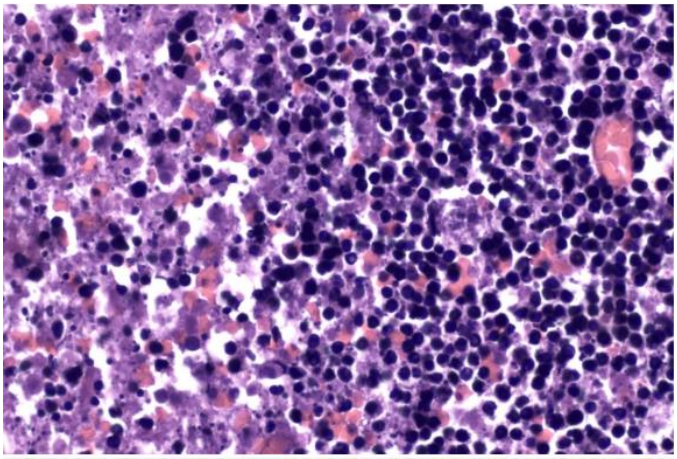

Figure 2: Necrotic debris with eosinophilic granular material.

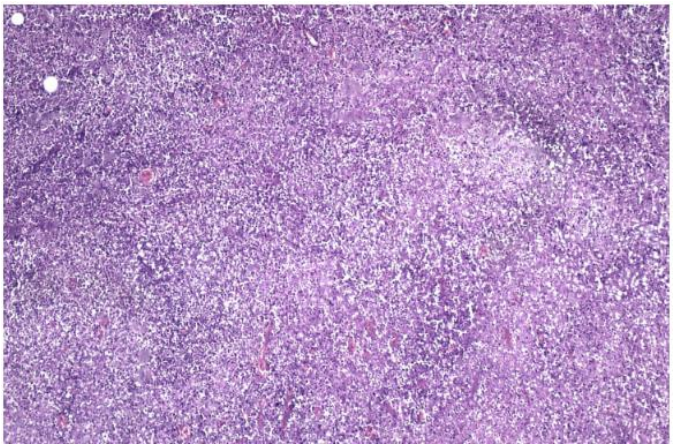

Figure 3: Distortion of normal lymph node architecture.

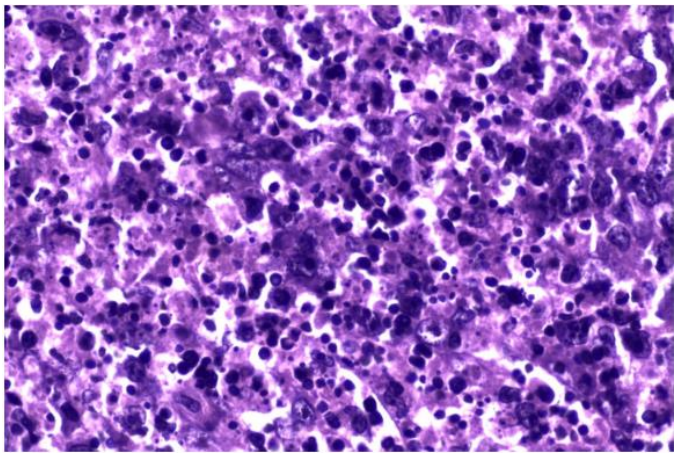

Figure 4: Histiocytes with admixed lymphocytes and nuclear debris.

Discussion

Epidemiology

Kikuchi-Fujimoto Syndrome (KFS) predominantly affects young women in their second to fourth decades of life. Though initially described in Japan, it has been reported worldwide. The incidence remains uncertain due to under diagnosis and misdiagnosis [4,5].

Pathogenesis

The exact cause of KFS remains unknown. Viral infections (e.g., Epstein-Barr virus, human herpesvirus 6 [HHV-6], cytomegalovirus [CMV]) and autoimmune mechanisms have been implicated. Some cases are associated with systemic lupus erythematosus (SLE), suggesting an immunological link [1,6].

Clinical Presentation

Patients typically present with acute or subacute cervical lymphadenopathy, low-grade fever, night sweats, and fatigue. Less common symptoms include weight loss, hepatosplenomegaly, and skin rashes. Leukopenia is a frequent hematological finding [1,5-7].

Differential Diagnosis

KFS must be distinguished from conditions such as:

- Tuberculosis: Acid-fast bacilli testing helps exclude tuberculous lymphadenitis.
- Lymphoma: Differentiation requires histopathological examination.
- Systemic Lupus Erythematosus (SLE): Autoantibody testing (ANA, anti-dsDNA) aids in differentiation.

- Other Infectious Causes: Viral serologies should be performed to rule out Epstein-Barr virus, CMV, and other infections [1,4,8,9].

Diagnosis

The gold standard for diagnosis is a lymph node biopsy, which reveals paracortical necrosis, karyorrhectic debris, and histiocytic infiltrates. Unlike infections or malignancies, neutrophils and plasma cells are absent.

Laboratory findings are usually nonspecific [1,2,9,10].

Management and Prognosis KFS is a self-limiting disease, with symptoms resolving within one to six months. Treatment is mainly symptomatic:

-Nonsteroidal anti-inflammatory drugs (NSAIDs) for fever and pain management.

- Corticosteroids in severe cases with extensive lymphadenopathy or systemic symptoms.

Close monitoring is required to rule out progression to SLE or recurrence of symptoms [1,2,4-7].

Conclusions

Kikuchi-Fujimoto Syndrome (KFS) is a rare and often under-recognized condition that should be considered in patients presenting with febrile lymphadenopathy. Accurate diagnosis through histopathological examination is essential to prevent unnecessary treatments for other conditions such as lymphoma. The disease follows a benign course with an excellent prognosis; however, long-term follow-up is recommended to monitor for potential autoimmune complications.

Future research should focus on improving the understanding of the immunological aspects of KFS and its overlap with autoimmune diseases. Additionally, the development of advanced imaging techniques may aid in differentiating KFS from malignancies. Genetic studies exploring susceptibility to KFS could provide further insights into its pathogenesis and potential risk factors.

Additional Information

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all 4 of 5participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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