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Kikuchi-Fujimoto Disease in a Five-Year-Old Female Presenting with Cervical Lymphadenopathy: A Rare Pediatric Case Report

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Abstract

Background: Kikuchi-Fujimoto disease (KFD), also known as histiocytic necrotizing lymphadenitis, is a rare, benign, self-limiting cause of cervical lymphadenopathy. Although commonly described in young adults, pediatric cases are uncommon and often mimic tuberculosis, lymphoma, and systemic lupus erythematosus (SLE).

Case Presentation: Ours is a 5-year-old female presented with unilateral cervical lymphadenopathy of two weeks duration. The swelling was associated with fever and tenderness. Laboratory investigations revealed mild anemia ,Mildly elevated ESR and C-reactive protein with Leukocytosis.

Fine-needle aspiration cytology suggested necrotizing lymphadenitis. Excisional lymph node biopsy demonstrated paracortical necrosis with abundant karyorrhectic debris, numerous crescentic histiocytes, plasmacytoid dendritic cells, and absence of neutrophils, confirming Kikuchi-Fujimoto disease. The patient received supportive treatment and showed complete clinical recovery during follow-up.

Conclusion: KFD should be considered in the differential diagnosis of persistent cervical lymphadenopathy in children. Histopathological examination remains the gold standard for diagnosis and helps avoid unnecessary antitubercular therapy or chemotherapy.

Keywords: Kikuchi-Fujimoto disease, Histiocytic necrotizing lymphadenitis, cervical lymphadenopathy, Pediatric pathology, Case report.

Introduction

Kikuchi-Fujimoto disease (KFD) is a rare benign necrotizing lymphadenitis first described independently by Kikuchi and Fujimoto in Japan in 1972. The disease predominantly affects young women of Asian origin, while pediatric cases are distinctly uncommon. Because the clinical presentation overlaps with tuberculosis, lymphoma, and autoimmune diseases, diagnosis is frequently delayed. Histopathological examination of an excised lymph node remains the diagnostic gold standard.

Case Presentation

A 5-year-old female child presented with a swelling on the left side of the neck for two weeks. The swelling gradually increased in size and was associated with mild pain and fever . There was no history

of chronic cough, weight loss, night sweats, recent upper respiratory tract infection, or tuberculosis contact.

General physical examination was unremarkable. Local examination revealed enlarged cervical lymph nodes measuring 2 × 1.5 cm, which were firm, mobile, and tender. No hepatosplenomegaly or generalized lymphadenopathy was noted.

Laboratory investigations showed:

- Hemoglobin- Anemia
- Total leukocyte count: mild elevated
- ESR: mild elevated
- CRP: mild elevated
- LDH: mild elevated

Tests for tuberculosis and autoimmune disorders were negative.

Ultrasonography of the neck demonstrated multiple enlarged cervical lymph nodes with preserved hilar architecture.

Fine-needle aspiration cytology showed necrotizing lymphadenitis with abundant karyorrhectic debris and histiocytes.

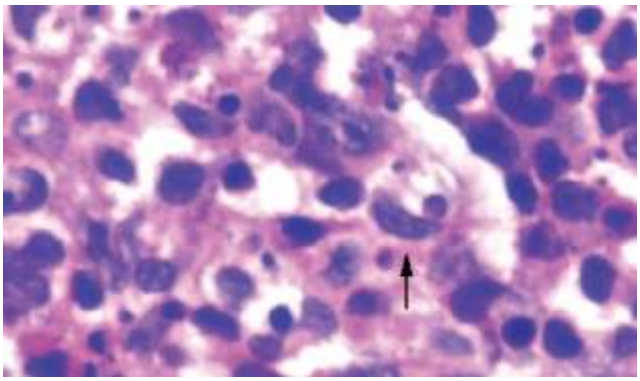

Figure 1; Cytology showing crescent shaped histiocytes.

An excisional lymph node biopsy was performed.

Histopathological Findings

Microscopic examination revealed preserved nodal architecture with patchy paracortical necrosis containing abundant apoptotic nuclear debris (karyorrhexis). Numerous crescent-shaped histiocytes and plasmacytoid dendritic cells surrounded the necrotic areas. Neutrophils and eosinophils were absent. No granulomas or malignant cells were identified. These findings were diagnostic of Kikuchi-Fujimoto disease.

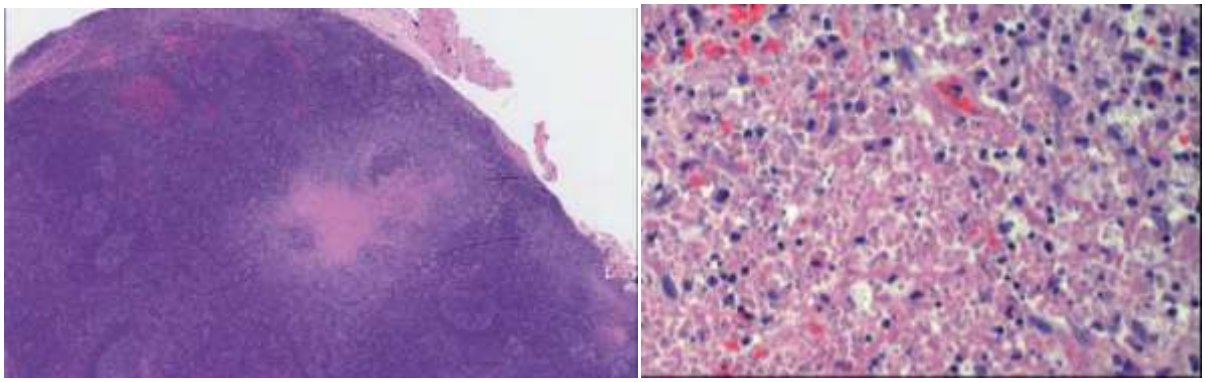

Figure 2 : Showing lymph node with Preserved architecture with follicular hyperplasia. Paracortex is expanded and shows areas of abundant karryorehtic debris and necrosis .

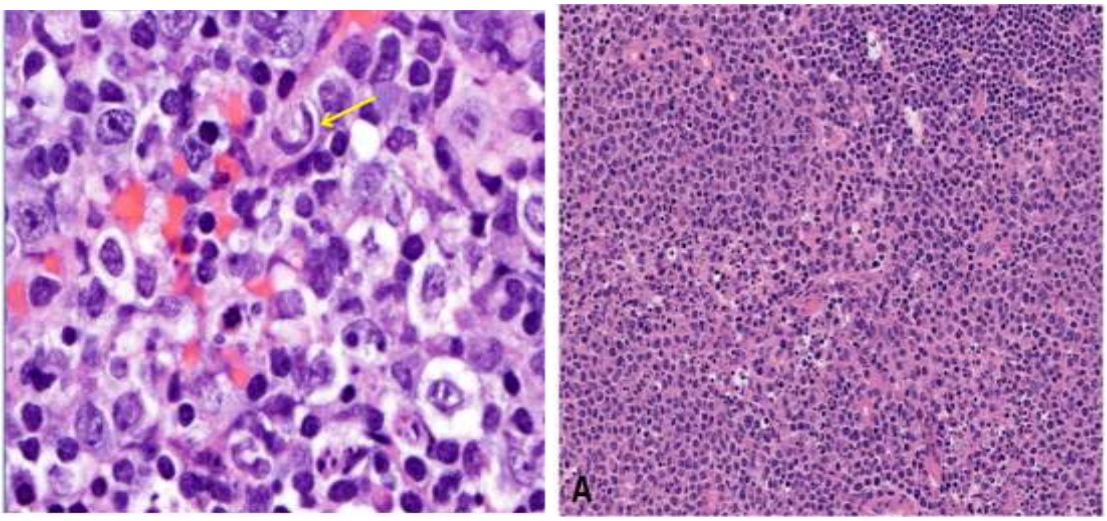

Figure 3: Histopathology showing crescent shaped histiocytes. Paracortical sheets of histiocytes with karyorrhectic debris

Immunohistochemistry, shows predominance of CD68-positive histiocytes and CD3-positive T lymphocytes, supporting the diagnosis.

The patient was treated conservatively with antipyretics and analgesics. Corticosteroids were not required. At follow-up after two weeks, cervical lymphadenopathy had resolved completely, and the child remained asymptomatic.

Discussion

KFD is an uncommon cause of cervical lymphadenopathy in childhood. Although the exact etiology remains uncertain, viral and autoimmune mechanisms have been proposed. Cervical lymphadenopathy with fever is the most common clinical presentation.

Epidemiology-First described in Japanese patients and has a higher prevalence among Asians. It usually affects young adults (younger than 40 years), but it can occur in any age group. Most reports show female predominance. However, some studies from Asian countries suggest that the male to female ratio is closer to 1:1.

Clinical features can vary from Self-limiting to evolving over several weeks. Posterior cervical lymphadenopathy is the most common site .The effected lymph nodes are Tender and painful.

Pathogenesis of Kikuchi disease is unknown. Based on the clinical presentation, course, and histologic changes

Hypothesis is that it could be due to one of these.

A. Infectious agents - Numerous inciting agents have been proposed .

- Human Herpes viruses
- Parvovirus B19
- Paramyxovirus
- HIV
- Toxoplasma gondi

B. Genetics

C. Autoimmune disease

Histologic and clinical changes suggest an immune response of T cells and histiocytes to an infectious agent. **However, no study has definitively proven a causal relationship between a virus and KFD or identified viral particles ultra structurally .**

Autoimmune cause-KFD shares sex and age predisposition as well as histologic features with systemic lupus erythematosus .Kikuchi disease reflects a self-limited, autoimmune condition like – SLE.Known to caused by virus-infected transformed lymphocytes.

Genetics cause- Exuberant T-cell–mediated immune response for Antigens - in genetically susceptible people.Human leukocyte antigen (HLA) class II alleles, specifically HLA-DPA1 and HLA-DPB1 - prevalent in Asians and are extremely rare or absent in whites.

Under Electron microsopy Patients of KFD and SLE show similar tubular reticular structures in lymphocytes and histiocytes. Apoptotic cells shows nuclear chromatin condensation ,Fragmentation along the nuclear membrane, Intact organelles and histiocytes phagocytosing karyorrhectic debris.

Serologic tests like Rheumatoid factor, Antinuclear antibodies, and Anti–double-strand DNA antibodies have been consistently negative in KFD patients.

The major differential diagnoses include tuberculous lymphadenitis, lymphoma, and lupus lymphadenitis. Histopathological examination is essential because clinical and radiological findings are nonspecific. Characteristic microscopic findings include patchy necrosis, abundant karyorrhectic debris, crescentic histiocytes, plasmacytoid dendritic cells, and absence of neutrophils.

Early tuberculosis vs KFD

- KFD and TB could have coexisted and TB got flared with steroids .
- Patient could have developed TB as a complication of steroids Induced immunosupression used for KFD treatment.
- The concomitant occurrence of tuberculosis and

KFD has been rarely reported in the literature.

SLE vs KFD

- Most difficult differential diagnosis to resolve.
- Histologically and IHC indistinguishable from KFD.

SLE vs KFD

- Interestingly there are rare documented associations between KFD and SLE .
- The disease could be unusual presentation of SLE.

- However, it is also possible that some of these cases are actually lupus lymphadenitis which are misdiagnosed as Kikuchi disease.
- **Serologic tests** - Rheumatoid factor, Antinuclear antibodies, and Anti–double-strand DNA antibodies have been negative in KFD patients.

Our patient represents a rare pediatric presentation. Early recognition prevented unnecessary antitubercular treatment and invasive investigations. The disease followed its typical benign, self-limiting course.

KFD is Self-limited disease that usually resolves within a few months. Symptomatic treatment is enough.No specific therapy for these patients. It has a low recurrence rate of 3% to 4%

Patients with relapsing disease or a more severe clinical course might benefit from corticosteroid therapy.

Conclusion

KFD is a Significant diagnostic challenge to pathologists and clinicians. Excisional lymph node biopsy is the optimal specimen for diagnosis of KFD.

IHC stains can be helpful to differentiate from lymphomas. From a clinical standpoint, one of the most important differential diagnoses is SLE, which should be ruled out in all patients diagnosed with KFD.

Although rare in children, Kikuchi-Fujimoto disease should be considered in the differential diagnosis of persistent cervical lymphadenopathy. Excisional lymph node biopsy remains the gold standard for diagnosis. Prompt recognition avoids misdiagnosis and inappropriate therapy.

Patient Consent

Written informed consent was obtained from the patient's parent/legal guardian for publication of this case report and accompanying images.

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