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A RARE CASE OF KIKUCHI-FUJIMOTO DISEASE IN A 28-YEAR-OLD WOMAN PRESENTING AS CERVICAL LYMPHADENITIS: A CASE REPORT AND LITERATURE REVIEW

Surgery

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ABSTRACT

Kikuchi-Fujimoto disease (KFD), or histiocytic necrotizing lymphadenitis, is a rare, self-limiting condition primarily affecting young women. It is characterized by cervical lymphadenopathy, fever, and systemic symptoms that can mimic other diseases, including lymphoma and systemic lupus erythematosus (SLE).We report a case of a 28-year-old woman who presented with cervical lymphadenitis, fever, and fatigue. Initial investigations, including imaging and blood tests, suggested an infectious or neoplastic process. However, a lymph node biopsy confirmed Kikuchi-Fujimoto disease. The patient was managed conservatively, and her symptoms resolved within 3 months.KFD is a diagnostic challenge due to its overlapping clinical features with other serious conditions. The exact pathogenesis remains unclear, though viral and autoimmune triggers are suspected. Review of the literature reveals the importance of distinguishing KFD from more aggressive diseases. KFD should be considered in the differential diagnosis of cervical lymphadenopathy, especially in young women. Accurate diagnosis is crucial to avoid unnecessary aggressive treatments.

KEYWORDS

Kikuchi-Fujimoto disease, histiocytic necrotizing lymphadenitis, cervical lymphadenopathy, young woman, biopsy, self-limiting.

INTRODUCTION

Kikuchi-Fujimoto disease (KFD), first described independently by Kikuchi and Fujimoto in 1972, is a rare, benign, self-limiting condition characterized by regional lymphadenopathy, most commonly involving the cervical nodes. It typically affects young adults, with a female predominance, and presents with nonspecific symptoms like fever, fatigue, and night sweats [1]. Due to its rarity and clinical overlap with other disorders such as lymphoma, tuberculosis, and autoimmune diseases like systemic lupus erythematosus (SLE), KFD is often misdiagnosed.Histopathologically, KFD is characterized by necrotizing lymphadenitis, with the absence of neutrophils, a hallmark that helps distinguish it from other conditions[2]. Although the etiology of KFD remains uncertain, infectious (particularly viral) and autoimmune mechanisms have been proposed. This case report details the clinical presentation, diagnosis, and management of a 28-year-old woman with Kikuchi-Fujimoto disease, presenting as cervical lymphadenitis, and provides a review of the literature to highlight the importance of accurate diagnosis and management.

Case Report

A 28-year-old previously healthy woman presented to the outpatient clinic with a 3-week history of progressively enlarging, tender cervical lymph nodes on the left side, associated with low-grade fever, night sweats, and malaise. She reported no significant weight loss, cough, or history of recent infections. There was no family history of autoimmune disorders, malignancy, or tuberculosis. On physical examination, multiple tender, mobile, firm lymph nodes, approximately 1-2 cm in diameter, were palpated in the left cervical region.Initial laboratory investigations revealed a mild leukopenia with a white blood cell count of 3,500/μL, and elevated erythrocyte sedimentation rate (ESR) at 45 mm/hour. Blood cultures were negative, and tests for Epstein-Barr virus (EBV), cytomegalovirus (CMV), and HIV were unremarkable. Chest X-ray showed no abnormalities, and a contrast-enhanced CT scan of the neck revealed multiple enlarged cervical lymph nodes without central necrosis or calcifications, raising suspicion for lymphoma or an infectious etiology. But serum LDH Levels and serum ADA were within normal range.

A fine needle aspiration biopsy was performed but yielded inconclusive results. A subsequent excisional biopsy of one of the affected lymph nodes revealed histiocytic necrotizing lymphadenitis, with abundant karyorrhectic debris, paracortical areas of coagulative necrosis, and the absence of neutrophils, findings consistent with Kikuchi-Fujimoto disease as shown in figure 1 and 2. Bcl2 marker was done which was reported to be negative on ihc as shown in figure 3.The patient was managed conservatively with non-steroidal anti-inflammatory drugs (NSAIDs) for pain and fever. The post operative wound site was healthy with no discharge. Given the self-limiting nature of KFD, no corticosteroids or antibiotics were administered. She was monitored closely, and over the following 3 months, her symptoms gradually resolved, with complete regression of the lymphadenopathy.

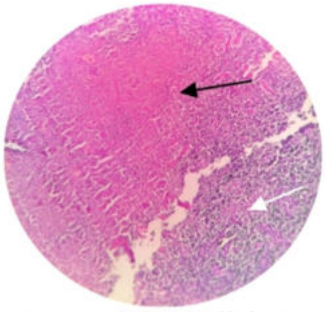

Figure 1magnified view with **black arrow** pointing towards area of necrosis and **white arrow** pointing towards area of inflammatory cells

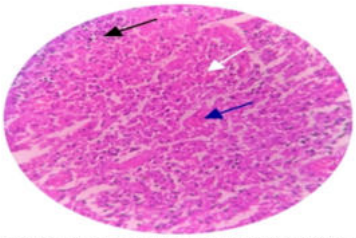

Figure 2 **Black arrow** showing area of lymphocytes and plasma cells,**white arrow** shows karyorrhectic debris and **blue arrow** shows dark granular eosinophilic necrosis.

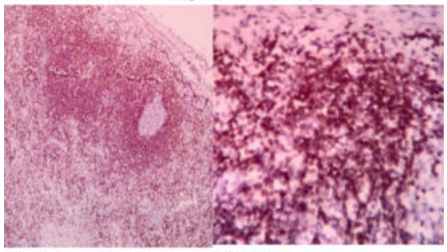

Figure 3 showing BCL2 marker negative in IHC of this patient.

DISCUSSION

Kikuchi-Fujimoto disease remains a diagnostic enigma due to its rare incidence and nonspecific presentation. It predominantly affects young women, with the majority of cases occurring between the ages of 20 and 40 [3]. The hallmark feature of KFD is cervical lymphadenopathy, which can be confused with malignant lymphomas, particularly Hodgkin's lymphoma, or infectious lymphadenopathies, such as tuberculosis. Fever, often persistent and low-grade, is present in most cases, and systemic symptoms like fatigue, night sweats, and weight loss may also occur, further complicating the diagnosis[4].

The definitive diagnosis of KFD is made through histopathological examination of affected lymph nodes. The characteristic findings include areas of necrosis with abundant apoptotic cells and histiocytes, but notably, the absence of neutrophils and granulomas helps to differentiate KFD from other necrotizing lymphadenopathies, such as tuberculosis and autoimmune disorders like lupus lymphadenitis. Although the pathogenesis of KFD is still debated, many researchers postulate that the disease represents an immune response to viral infections. Various viral triggers have been implicated, including Epstein-Barr virus (EBV), human herpesvirus 6 (HHV-6), and cytomegalovirus (CMV), though no specific pathogen has been definitively linked to the condition. Additionally, an association with systemic lupus erythematosus (SLE) has been suggested, with some reports indicating a potential overlap between the two diseases, particularly in individuals who later develop autoimmune diseases[5].Hence autoimmune disease workup was done in our patient including ANA profile, APLA,ANCA tests which turned out to be negative. KFD is generally a self-limiting condition, with most cases resolving within 1 to 4 months without the need for aggressive therapy. Treatment is primarily supportive, focusing on symptom relief with NSAIDs or corticosteroids in more severe cases. However, patients should be monitored for potential progression to or development of autoimmune diseases, particularly SLE, which can occur in a minority of cases. Given its benign course, it is crucial to avoid overtreatment, especially with cytotoxic or immunosuppressive therapies typically used for malignancies, highlighting the importance of accurate diagnosis. Review of available literature reveals that KFD is often misdiagnosed, leading to unnecessary investigations and treatments[6]. The disease is most prevalent in Asian populations but has been reported globally. Clinicians should maintain a high index of suspicion for KFD in young women presenting with cervical lymphadenopathy and systemic symptoms, particularly when initial infectious and neoplastic workup is inconclusive.

CONCLUSION

Kikuchi-Fujimoto disease is a rare, benign condition that should be considered in the differential diagnosis of cervical lymphadenopathy, particularly in young women. Early recognition and accurate diagnosis through histopathological analysis of lymph node tissue are crucial in preventing unnecessary treatments for suspected malignancy or infections. Although self-limiting, KFD requires careful follow-up to ensure resolution and to monitor for the potential development of autoimmune diseases like SLE.

Conflict Of Interest : None.

Ethical issues: None. Proper written consent from patient taken and patients identity is not disclosed.

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