International Journal Dental and Medical Sciences Research
Volume 4, Issue 6, Nov-Dec 2022 pp 699-701 www.ijdmsrjournal.com ISSN: 2582-6018

Kikuchi Fugimoto Disease with an unusual axillary lymphadenopathy: a Case Report

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Submitted: 08-12-2022 Accepted: 16-12-2022

ABSTRACT
Kikuchi-Fujimoto disease (KFD) is a benign and self-limited disorder, characterized by regional cervical lymphadenopathy with tenderness, usually accompanied with mild fever and night sweats. KFD is an extremely rare disease known to have a worldwide distribution with higher prevalence among Japanese and other Asiatic individuals.

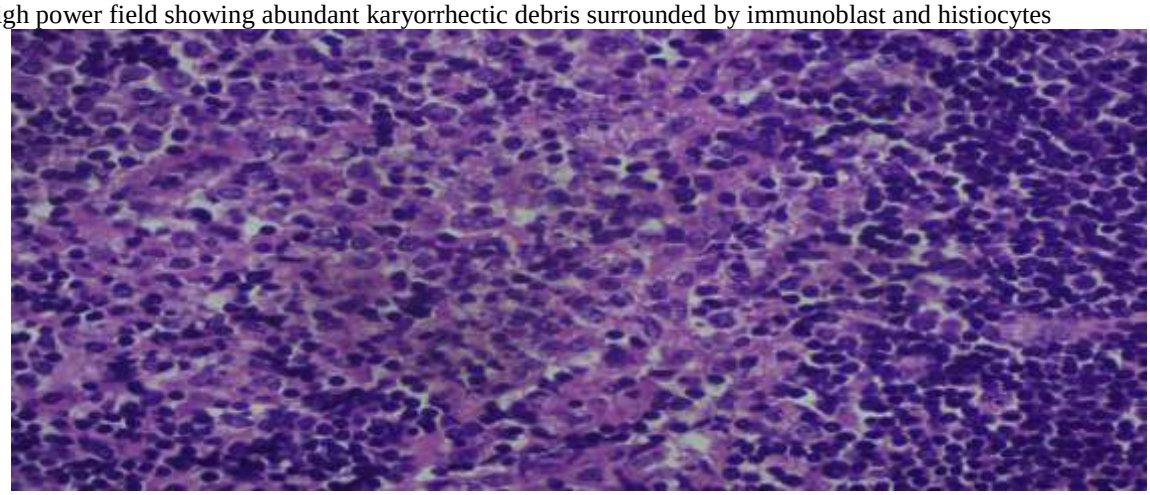
I. INTRODUCTION

KFD is a benign disease that presents clinically with cervical lymphadenitis and mild constitutional symptoms [(1).] However,involving axillary lymphadenitis as a presenting manifestation is rare, particularly when no other lymph node involvement are initially identified[(3).] We describe a young female, who presented with axillary lymphadenitis with history of fever and significant weight loss initially thought to be due to tuberculous lymphadenitis and later confirmed to have KFD on lymph node biopsy.

CASE DESCRIPTION

19 year old female presented with on and off fever and swelling in left axilla associated with pain since 2 weeks with significant weight loss of 9kg in 3 months and loss of appetite .On examination, only 1 left anterior axillary lymph node was palpable 2x1cm and was tender. No other lymph nodes palpable and systemic examination within normal limit. Blood parameters were within normal range except for an elevated ESR-40. Chest x ray was normal.Ultrasound abdomen did not show any sonological abnormalities. Peripheral smear was normal. Blood culture showed no growth.Serology was non reactive. On suspicion of tuberculosis (TB) ,USG of left axilla was done which suggestive of multiple enlarged lymph nodes with compressed fatty hilum in the left axilla - likely infective etiology . However excision biopsy was done and also sent for CBNAAT. Patient was managed with IV antibiotics, fluids and other supportive measures .Patient improved symptomatically and discharged. After 1 week, CBNAAT was negative for TB and histopathology report suggestive of - HISTIOCYTIC NECROTIZING LYMPHADENITIS→KIKUCHI FUJIMOTO DISEASE. Patient was managed with short course of NSAID and steroids. Patient improved thereafter with no further fever spikes and swelling subsided.

High power field showing abundant karyorrhectic debris surrounded by immunoblast and histiocytes



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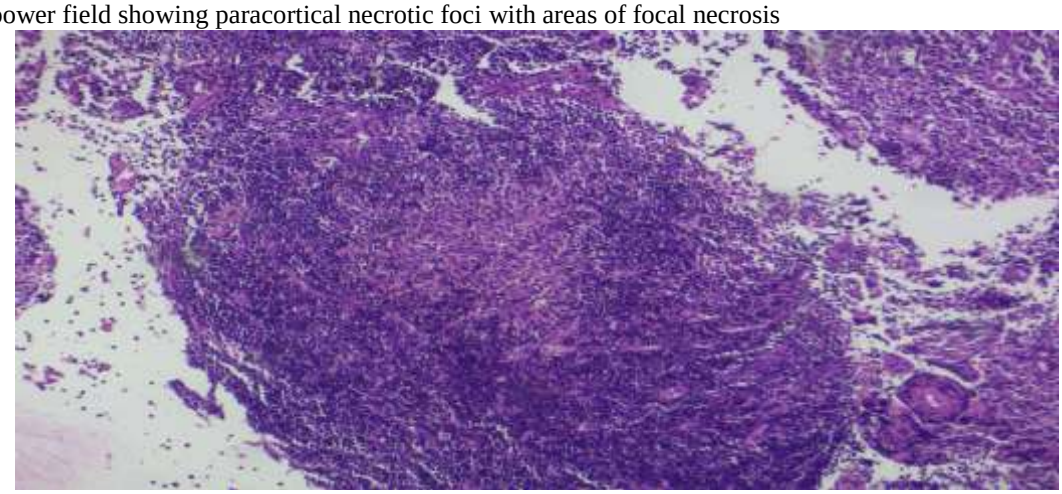
Low power field showing paracortical necrotic foci with areas of focal necrosis



II. DISCUSSION

Kikuchi disease (KD), also known as Kikuchi-Fujimoto disease or histiocytic necrotizing lymphadenitis, is a rare disease with an unknown cause that was first described in Japan in 1972 by Kikuchi and Fujimoto[(2).]The hallmarks of KD are prolonged fever and cervical lymphadenopathy. Skin rash, nausea, vomiting, fatigue, weight loss, joint pain, and sore throat are also common symptoms. It is more common in young Asian females, and only a few cases have been reported in Europe. The aetiology of the disease has been proposed using viral and immunological theories. Laboratory results typically revealed cytopenia and atypical lymphocytosis. It has a connection with SLE.The lymphadenopathy is typically cervical, with nonspecific symptoms such as fever and night sweats, making the diagnosis more difficult because the differential diagnosis is broad.[(4)]

Kikuchi's disease must be considered in the differential diagnosis of lymph node enlargement, along with SLE-associated lymphadenitis, malignant lymphoma, tuberculosis, herpes simplex lymphadenitis, plasmacytoid T cell leukaemia, Kawasaki's disease, acute myeloid leukaemia, infectious mononucleosis, sarcoidosis, and metastatic adenocarcinoma. [(6)]The majority of patients had a benign and self-limited course with spontaneous remission after weeks to months of illness. Early detection of KD may help to avoid unnecessary diagnostic and therapeutic interventions.

Our patient presented with axillary lymphadenitis and weight loss which was more suspicious of tuberculosis ,however CBNAAT was negative for TB and prompt analysis of histopathological study led to an definitive diagnosis of KFD.Therefore ,diagnosis is based on the characteristic histopathological features of the lymph node biopsy that shows paracortical necrotic foci with abundant karyorrhectic debris surrounded by immunoblast and histiocytes.Histologically it differs from tuberculosis by the classic biopsy features and the absence of caseatinggranulomas[(5).]

In patients with KD, histiocytic necrotizing lymphadenopathy is most commonly found in the cervical region. Lymph nodes (LNs) in extra-cervical body sites have been shown to be affected. However, the frequency and location of LN involvement varied significantly across reports. In a series of 244 patients, for example, Kucukardali reported a small percentage (5%) of generalised LNs involvement. Rimar found retroperitoneal LNs and generalised lymphadenopathy in 21% and 26% of 19 KD patients, respectively.The data on the detailed distribution of lymphadenopathy in patients with KD were sparse, and the reported incidences may be underestimated because extra-cervical images were not routinely performed in patients with clinical suspicion of KD[(4).] Kikuchi's disease is usually treated with NSAIDs to relieve lymph node tenderness and fever. Our patient improved after receiving symptomatic treatment. Corticosteroids are prescribed for severe cases of the disease. The disease is usually self-limiting andspontaneous recovery occurs in 1 to 4 months. It reoccurs at a rate of 3-4%. Patients with Kikuchi-Fujimoto disease should be followed-up for several years to survey the possibility of the development of systemic lupus erythematosus.[(1)]

III. CONCLUSION

Our patient presented with axillary lymphadenitis which is the intriguing case because of its uncommon presentation for KFD which prompts investigation and recognition by physician

and pathologist to avoid misdiagnosing it as TB or lymphoma and avoid inappropriate treatment.

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