KIKUCHI-FUJIMOTO DISEASE (KFD): A Rare Case Report

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Abstract— *Kikuchi-Fujimoto disease (KFD) is a rare disease. It has a worldwide distribution with a higher prevalence in Asians. KFD is usually a self limiting disease and benign in nature. Clinically it presents as regional cervical lymphadenopathy and sometimes may presents as generalized lymphadenopathy as well. Night sweats and low grade fever may also be associated in some cases. A case of a 36 year old female had attended in SMS Hospital. She presented with fever, weight loss and tender cervical lymph nodes. Diagnostic significance of KFD lies in that it may mimic some of the common diagnosis like SLE (Systemic Lupus Erythematosis), Tuberculosis, lymphoma and rarely adenocarcinoma. Diagnosis is usually made by biopsy of lymph node which shows fragmentation, necrosis and karyorrhexis. On thorough investigation this case was found of Kikuchi-Fujimoto disease (KFD) which is a rare disease. So case presenting with fever, weight loss and tender cervical lymph nodes should also be investigated for Kikuchi-Fujimoto disease (KFD).*

Keywords: Kikuchi-Fujimoto disease (KFD).

I. INTRODUCTION

Cervical lymphadenopathy is a common manifestation of varied etiology. The illness ranges from benign infectious etiology to malignant lymphoma.

KFD is also known as histiocytic necrotizing lymphadenitis. It was first described by Kikuchi and Fujimoto in 1972 so called Kikuchi-Fujimoto disease (KFD). It is a rare, benign and self limited disorder of unknown cause.

It is characterized by tender cervical lymphadenopathy, constitutional symptoms such as fever and night sweats[1]. Patient of KFD is usually younger with mean age of 21 years[2]. It has a slight female preponderance[3]. It is a self limiting disease with no specific therapy.

It may be an under-diagnosed condition and as it has an excellent prognosis. It should be differentiated from malignant lymphomas. Its knowledge should be there in both clinicians as well as pathologists who will help in preventing misdiagnosis and inappropriate treatment[1]. Recurrence is around 4%. It should always be considered in differential diagnosis of patients with younger age, cervical lymphadenopathy and biopsy features of necrosis, fragmentation and karyorrhexis[4, 5, 6].

II. METHODOLOGY

A rare case of Kikuchi-Fujimoto disease (KFD) who was presented with fever, weight loss and tender cervical lymph nodes at SMS Medical College, Jaipur (Rajasthan) India. On investigation it came out to Kikuchi-Fujimoto disease (KFD) which is a very rare disease. So case was studied thoroughly and case report was prepared to publish this rare case.

III. CASE REPORT

A case of 36 year old female presenting to with left sided neck swelling for 1 month. Swelling was associated with pain from the beginning. Patient had a history of fever for last 1 month. Fever was high

grade with chills and rigor at that time. There was no associated sore throat, burning micturition or abdominal pain. Patient also had a history of night sweats and weight loss for the last one month. There was no history of joint pains. She also complained of similar swelling in left axillary region for 7 days which regressed on its own. Patient was non-smoker, non-alcoholic. There was no history of insect bite, any chronic drug abuse; contact with animals etc.

On examination patient was febrile with a temperature of 102.4°F. There was a chain of tender lymph nodes extending from just below the left ear to left supra-clavicular region. The size of these nodes ranges from 2 to 4 cms. There were no palpable nodes on right side. These nodes were tender and mobile.

On routine investigation, CBC, TLC, LFT, RFT were found to be normal. USG neck revealed enlarged cervical lymph nodes on level 2, 3 and 4 showing hypo-echogenicity, necrotic foci and matting with maintained hilar vascularity and a diagnosis of necrotic cervical lymphadenopathy was kept. Bone marrow examination was done which was normal. Biopsy showed lymph nodal tissue with paracortical, well circumscribed necrotic lesions with karyorrhectic debris and fibrin deposits. There were numerous plasmacytoid monocytes, phagocytes and foamy histiocytes.

No plasma cells, neutrophills, no follicular hyperplasia, no atypia was found. AFB and GMS stains were negative for fungal microorganisms.

A diagnosis of histiocytic necrotizing lymphadenitis or KIKUCHI-FUJIMOTO DISEASE was established.

IV. DISCUSSION

KFD is benign histiocytic necrotizing lymphadenitis. It is a rare disease. KFD is most commonly present in Asians. Kikuchi and Fujimoto described KFD in Japan in 1972.[7, 8] Its causative agent has not been identified, although it is believed to be viral in origin. EBV, HHV 6 AND HHV 8 have been implicated. Autoimmune etiology has also been linked. It is associated with SLE at times. It tends to affect younger population below 30 years, mostly females but there are reported cases in older age group and pregnancy too.[9] As the disease is common in Asians, it has been linked to certain HLA alleles such as HLA Π, HLA-DPA 1 AND HLA-DPB 1. The most of KFD cases may be acute or subacute, usually evolves over 2-3 weeks.

The hallmark feature of KFD is unilateral lymphadenopathy with cervical involvement in 70-98% of cases.[2, 4, 10] However any lymph node region can be involved including axillary (14%) and supraclavicular (12%). Involved lymph nodes are small and mobile.

Fever can be the first symptom in 30-50% of cases.[4] Other symptoms are weight loss, nausea, vomiting, sore throat and night sweats. Skin lesions like maculopapular, mobiliform, urticarial rashes have been reported. There is an increased ESR and neutropenia.

A few patients have atypical lymphocytes in peripheral blood film[4] and sometimes, alkaline phosphatase and alanine phosphatase is increased.[1, 3, 4] Some patients with KFD have positive lab test for SLE and later they develop clinical SLE. The lack of monoclonal lymphocyte receptor rules out the possibility of lymphoma[4].

The differential diagnosis of a neck mass is extensive and includes lymphoma, metastasis, infectious mononucleosis, AIDS, cat-scratch disease, SLE, tuberculosis and others. Thus KFD is a rare differential of a commoner presentation and it should always be kept in mind when assessing neck nodes.

V. CONCLUSION

KFD is uncommon, but should feature in the list of differential diagnosis of tender cervical lymphadenopathy as its treatment is conservative and differs significantly from the other conditions that would be on that list such as TB, lymphoma, metastasis and SLE. Lymph node biopsy will aid in accurate diagnosis of necrotizing histiocytic lymphadenitis or Kikuchi-Fujimoto Disease.

CONFLICT OF INTEREST

None declared till now.

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