

Case Report

## **Kikuchi Fujimoto Disease presenting as Axillary Mass: A Case Report and Review of Literature**

**Gunjan Agarwal<sup>1</sup>, Hemant Jain<sup>2</sup>, Pankaj choudhary<sup>3</sup>, Amit Jain<sup>4</sup>**

<sup>1</sup>Consultant (EHCC hospital), <sup>2,3</sup>Resident, <sup>4</sup>Associate Professor,

Department of general surgery, Sawai Man Singh Medical College Jaipur – 302004, Rajasthan, India

**\*Corresponding author**

Dr. Gunjan Agarwal

Email: drgunjanagarwal80a@yahoo.co.in

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**Abstract:** Kikuchi Fujimoto disease is benign necrotizing histiocytic lymphadenitis. We report a case of 27 years old Indian female who came to us with right axillary mass and constitutional symptoms for two months. Excisional biopsy was done, which showed histological features of kikuchi fujimoto disease. KFD is an uncommon, under diagnosed disease, which is self-limiting in nature and has excellent prognosis. Two factors, infectious and immunological, are suggested to being responsible for this disease. Lymphoma and tuberculosis should be excluded first before making KFD as diagnosis and this can be possible only with the help of excisional biopsy of involved lymphnode. So KFD is the disease of exclusion, first exclude all possible cause of necrotizing lymphadenitis and then think about this disease. For Mangement, only supportive care is required but in severe cases steroid can be used. Some rheumatological diseases such as Systemic lupus erythematosus, Sjogren's syndrome may be associated with KFD.

**Keywords:** Kikuchi Fujimoto disease, Sjogren's syndrome

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**INTRODUCTION:**

Histiocytic Necrotizing lymphadenitis also known as Kikuchi-Fujimoto disease is first described by two pathologists, Kikuchi and Fujimoto in japan independently [1, 2]. Kikuchi and Fujimoto were doing study of patients treated for lymphoma. Some of them showed faster recovery then other, these patient actually had KFD. It is rare disease, benign in course and characterised by constitutional symptoms and localized lymphadenopathy [3]. Clinical manifestations of KFD are regional lymph-adenopathy which is subacute in nature, fever, skin rashes, leukopenia, neutropenia, and anemia, raised ESR and raised C-reactive protein. Microscopic study reveals that affected lymph node shows cortical and Para cortical necrosis. For its diagnosis, one should go for biopsy only because no specific imaging or clinical test is available for diagnosis of KFD. This disease mainly present in young Asian female. KFD also has tendency to recurrence in about 4 to 5% cases. NO specific treatment is required; it regresses spontaneously with an excellent prognosis. We describe a case of KFD IN 27 year Indian female who presented with multiple axillary lymph nodes with a short review of literature.

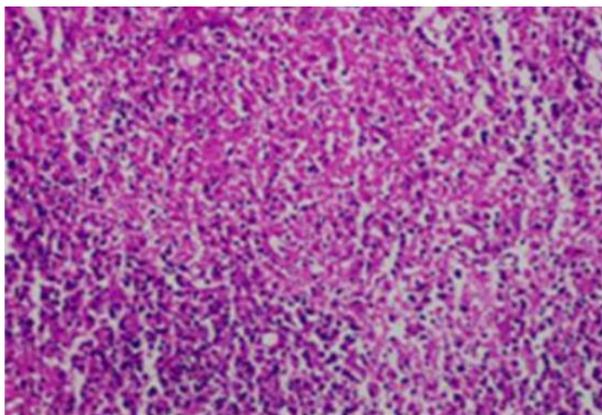
**CASE REPORT:**

A 27 years old female with no previous medical comorbidities presented to gynecology department with complain of right axillary swelling. Patient also complained of fever, anorexia, fatigue and pain. No history of alcohol, drug abuse, tobacco and medications. There was no significant past, personal and family history. Patient was fit and asymptomatic previously. No history of tuberculosis was there.

Patient was refered to Surgical OPD. In our department, proper examination was done. Significant findings were multiple palpable mobile axillary lymph nodes which were tender on touch and firm in consistency. She was lethargic, febrile, haemodynamically stable and well built. There was no pallor, hepato-splenomegaly and icterus. Blood test revealed neutropenia; raise ESR and C relative proteins. ECG, USG abdomen and Chest X-ray were grossly normal. Blood culture, urine culture, sputum culture were done. All came negative. Sputum culture was negative for AFB. Viral and auto immune screening were also done. Anti-nuclear antibody, rheumatoid factor, anti DNA antibody, HIV, Hepatitis B and C were negative.

She was treated symptomatically with anti-pyretic drugs and iv fluids. Lymph-adenopathy was persistent along with intermittent fever. She experienced improvement in her fatigue and malaise. USG of Right axilla showed multiple enlarged lymph nodes with fatty hilum and increase vascularity in right axilla s/o inflammatory lymphadenopathy. FNAC was done, which came inconclusive. Excisional biopsy was

done which showed lymph node tissue with Paracortical, well circumscribed necrotic lesion with karyorrhectic debris and fibrin deposits. There were numerous plasmacytoid monocytes, phagocytic and foamy histiocytes. No plasma cells, no neutrophils, no follicular hyperplasia, no atypia. AFB and GMS stains are negative for fungi and microorganisms. On the basis of microscopic findings diagnosis was confirmed.



**Fig 1: Lymph node showing multiple areas of necrotic foci with apoptotic debris.**

Patient was discharged after one week of inpatient stay. At follow up she had no symptoms and no abnormalities on clinical examination.

#### **DISCUSSION:**

Masahiro Kikuchi and Fujimoto, both first described it as lymphadenitis with focal proliferation of reticular cells with histiocytes and nuclear debris in separate journals [1, 2]. Aetiology has not been fully determined but probably there are two main factors responsible for KFD: 1) infection and 2) auto immune disorder.

Several infectious agents mainly Epstein Barr virus, Human herpes virus 6 and 8, Rubella, Parainfluenza, HIV, HTLV1, Dengue virus, Parvo virus B 19, Yersinia enterocolitica, Bartonella, Brucella, Toxoplasma etc have been suggested as etiological factor but none have been confirmed. Viral aetiology is supported by its nonspecific self-resolving symptoms, which are slow and insidious onset. Some people have hypothesized a possible link between SLE and KFD. But ANA, RF and other immunological parameters are not related to KFD. There is also evidence of genetic susceptibility for KFD. HLA class II genes (HLA-DPA1, HLA-DPB1) are more common found in that person who suffered from KFD and these genes are more commonly present in Asian population [11].

KFD is more commonly seen in females (male: female = 1:4), young adult (mean age of onset is 25) and Asian countries. Onset of disease is usually subacute. First symptom is usually fever in 70% cases [4]. Lymph-adenopathy is present which is <3cm in size, mobile, painless and tender some times. Other signs and symptoms include nausea, night sweating, malaise, anorexia, weight loss, leucopenia, diarrhoea and skin rashes [5]. Imaging (CT scan, MRI, and PET scan) not only provides inconclusive results but also creating confusion by mimicking of findings in lymphoma, metastasis and tuberculosis in which nodal necrosis is seen.

Most common affected lymph nodes are cervical (70%) specially jugular and posterior cervical chain, then axillary (14%) and supra-clavicular lymph nodes (12%). Rarely inguinal, thoracic and mesenteric lymph nodes are also affected. Skin lesions are present in 30% cases. Nonspecific, erythematous papules, plaque, acneiform and morbilliform lesion can be there in KFD. Mucosal involvement can be present. Skin biopsy finding is presence of lymphohistiocytes with karyorrhectic debris [7]. ESR is high in 70% cases of KFD [8].

Kwon et al found following findings on CT scan of patients suffering from KFD: (a) Multiple homogeneous lymph-adenopathy II to V groups, (b) In

94% case, size of lymph nodes are < 2.5 cm and (c) Perinodal infiltration and necrosis [12]. USG scans show lymph node with hypo-echoic center and hyper-echoic rim in case of KFD [13]. Definitive diagnosis is made by biopsy. FNAC has 56% accuracy [8]. Histo-pathological studies show occurrence of confluent necrotic area surrounded by cluster of lymphocytes, histiocytes, immunoblast, plasma cell but absence of neutrophil [9].

Microscopically findings in KFD show three patterns:

- 1) Proliferative- seen in 33% cases, dominant inflammatory infiltrates (histiocytes, plasmacytoid monocytes, lymphoid cell containing karyorrhectic nuclear fragments and eosinophilic apoptotic debris.
- 2) Necrotizing- seen in 50% cases, showing coagulative necrosis.
- 3) Xanthomatous- Rare, no granulocytes, only foamy macrophages present.

Immuno-histochemistry marker for diagnosis of KFD is CD68. Absence of monoclonal lymphocytes receptor rules out the possibility of lymphoma. Positive immuno-staining results by monoclonal antibody ki-MIP are seen in kikuchi disease, not in lymphoma. Association between SLE and KFD are reported but not well defined, share same age and sex predisposition and both have similar histological features. But in SLE, Azzopardi phenomenon and hematoxylin bodies present which are absent in KFD [10].

Differential diagnosis includes SLE, lymphoma, tuberculosis, sarcoidosis, syphilis, still's disease, cat scratch disease, infectious mononucleosis, viral lymphadenitis, Kawasaki disease etc. Only symptomatic treatment and reassurance are required. Spontaneous regression is observed within one to six months. Steroid like prednisolone, ciprofloxacin and minocycline can be used in severe disease but their use is not well established. Long term follow up of these patients are necessary because recurrent cases of KFD is reported.

#### CONCLUSION:

The aetiology, pathology, diagnosis and management of KFD are still mystery, not fully understood and matter of debate. Kikuchi Fujimoto disease is rare, benign, self-limiting disease with excellent prognosis but should be kept in list of differential diagnosis of lymph-adenopathy especially when it is affecting cervical chain of lymph nodes. For accurate diagnosis we should go for excisional biopsy. If there is confusion with SLE occurs, r/o SLE with the

help of immunological test. Once diagnosis of KFD has made, reassure and inform the patient about its benign course and self-limiting nature. Make sure that patient should come for follow up on OPD basis because the risk of associated rheumatological disease and recurrence of disease.

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