

## REVIEW ARTICLE

### KIKUCHI-FUJIMOTO DISEASE: A RARE CASE REPRESENTATION AND REVIEW OF LITERATURE

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**ABSTRACT:** It is also known as Kikuchi disease (Cervical subacute Necrotising Lymphadenitis) or Kikuchi histocytic necrotizing Lymphadenitis, it was first described in Japan by Dr. Masahiro Kikuchi in 1972<sup>(1)</sup> and independently by Y. Fujimoto. K.F.D is a rare, self-limiting disorder that typically affects the cervical lymph nodes, recognition of this condition is crucial, because it can easily be mistaken for Tuberculosis, or Lymphoma, awareness of this disorder will help to prevent misdiagnosis and inappropriate treatment<sup>(2)</sup>. We are here presenting a case of Kikuchi disease in 16 years old female diagnosed by excision biopsy and treated conservatively.

**KEYWORDS:** Cervical Lymphadenopathy, Kikuchi disease, Red Flag referral, histocytic necrotizing lymphadenitis.

**INTRODUCTION:** Persistent cervical lymphadenopathy is an important "Red Flag" referral criterion to head & neck clinics in adults over the age of 40 years, when lesions of Thyroid and salivary glands are excluded the majority (75%) of neck lumps are malignant. Kikuchi disease is a rare and a self-limiting illness, usually characterized by cervical lymphadenopathy and fever, which has symptoms which may overlap with Hodgkin's lymphoma or Tuberculosis leading to misdiagnosis in some patients, ANA, APCA, Anti-ds DNA, RF is usually negative and may help in differentiating from SLE. KD is mainly seen in Japan, Isolated cases are reported in America, Europe, and Asia. It is mainly a disease of young adults (20-30years), with a slight bias towards females. The cause of this disease is not known although infectious and autoimmune etiologies have been proposed, recurrence rate is about 3%, course of the disease is general benign and self-limiting, lymphadenopathy most often resolves over several weeks to 06months, mortality is extremely rare.

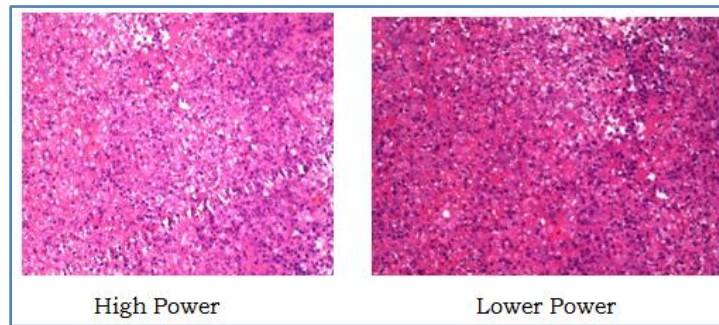
**CASE REPORT:** A 16 year old female came to ENT OPD with complaints of fever and swelling in left side of the neck (Posterior Triangle) since 15 days, she attended to general practitioner for above complaints treated for 02 weeks not getting improvement, she approached ENT specialist, initially treated with antibiotics and anti-inflammatory drugs, on examination she was febrile and swelling in the left side of the neck in the posterior triangle, on palpation 3 to 4 tender, matted palpable lymph nodes present in the left side of the neck. The patient was investigated to rule-out tuberculosis (Since the lymph nodes are matted) all the investigations were within normal limits, FNAC shows non-specific lymphadenitis advised excision biopsy.

Subsequently patient was taken up for excision biopsy under local anesthesia, a small horizontal incision is given over the swelling in the left side of the neck, one lymph node size 2cm x 1cm was picked up and sent for HPE the report came as Kikuchi's Lymphadenitis.

**Biopsy Report:** (11931/2014, Fig No.1) Section shows multiple bits of lymph node with follicles containing active germinal centers. Paracortex shows histocytes. There are foci of necrosis surrounded by histiocytes and neutrophils. No epithelioid cells seen. No giant cells noted.

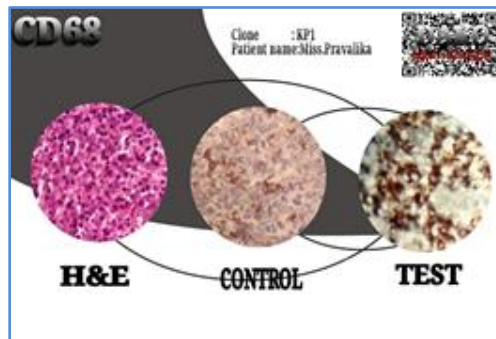
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**Impression:** Kikuchi's Lymphadenitis, advised for IHC studies for CD 68.



**Fig. 1: HPE Report**

**I.H.C. Report:** (Block No 1931/5028). Histiocytes are positive for CD 68. (Figure No.2)



**Fig. 2: I.H.C. Report**

After 02 days Patient was discharged under antibiotic coverage, suture removal done on 7<sup>th</sup> day, wound was healthy patient is doing well she has been referred to physician for further management.

**DISCUSSION:** Kikuchi disease is a benign disorder of unknown cause with characteristic clinical findings and affected lymph nodes showing lesions with Karyorrhectic nuclear debris and proliferating mononuclear cells, including histiocytes, lymphoid cells and Plasmacytoid T cells/Plasmacytoid monocytes (CD68+), KFD was first described in 1972 by Kikuchi and Fujimoto. In 1982 the first cases of KFD were reported in North America and Europe<sup>(3)</sup> and the disease is now recognized worldwide, KFD commonly affects young women with a peak age of incidence occurring in the third decade<sup>(4)</sup> but rarely affects patients under 16 year of age. The ratio of disease occurrence between females and males is 4:1, although less apparent gender differences occur in Asian population.

The etiology of KFD remains unclear. Several infective agents including EBV, parvovirus B19 and HHV-6<sup>(5)</sup>, have been postulated as causative although no relationship has yet been established, due to the similar histology seen in KFD and the lymphadenitis of SLE and that both diseases most often occur in young females, Dorfman and Berry<sup>(6)</sup> suggested KFD could be an attenuated form of SLE. Another explanation<sup>(7)</sup> proposed that KFD might be a self-limiting SLE-like autoimmune reaction to viral infected transformed lymphocytes. Whatever the pathogenesis, it is likely that the

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development of KFD is a multifactorial process involving environmental, biological and genetic influences.

The signs and symptoms of Kikuchi's disease are fever, lymphadenopathy, skin rashes and headache. Rarely hepatosplenomegaly and nervous system involvement resembling meningitis is seen. Differential diagnosis includes SLE, disseminated tuberculosis, Lymphoma, sarcoidosis, and viral lymphadenitis. The histopathological features of KFD are quite distinctive and the only mimic is SLE lymphadenitis. The lymph node changes in Kawasaki disease (Mucocutaneous lymph node disease), cat scratch disease and atypical mycobacterial infection are quite different, being characterized by intravascular fibrin thrombi and neutrophils, stellate micro abscesses with neutrophils and necrotizing stellate granulomatous inflammation respectively,<sup>(8)</sup> it is diagnosed by lymph node excision biopsy.

The majority of patients with KFD present with cervical lymphadenopathy usually of 1-4cm in diameter, the posterior cervical triangle being the most commonly affected site<sup>(9)</sup>, other less common signs and symptoms include splenomegaly, fever and weight loss – one third of patients have a rash at presentation, findings that can heighten the clinical overlap with infectious mononucleosis, SLE and lymphoma. A number of non-specific haematological abnormalities may occur in KFD. Approximately 50% of cases show a mild neutropenia<sup>(10)</sup> with leucopaenia also present in 25-40% of cases. Other non-specific findings include a raised CRP and ESR but their absence does not exclude KFD.

**TREATMENT MEDICAL CARE:** Treatment of Kikuchi's disease is generally supportive. Non-steroidal anti-inflammatory drugs (NSAIDs) may be used to alleviate lymph node tenderness and fever. The use of corticosteroids, such as prednisone, has been recommended in severe extra nodal or generalized Kikuchi's disease,<sup>(11)</sup> indications for corticosteroid use include the following:

1. Neurologic involvement:
  - Aseptic meningitis.
  - Cerebellar ataxia.
2. Hepatic involvement – Elevated LDH level.
3. Severe lupus like syndrome - Positive ANA titers.

Jang and colleagues recommended expanding the indications for corticosteroid use to less severe disease<sup>(12)</sup> they administered prednisone when patients had prolonged fever and annoying symptoms lasting more than 02 weeks despite NSAID therapy, as well as for recurrent disease and for patients who desired a faster return to work. Immunosuppressants have been recommended as an adjunct to corticosteroids in severe, life-threatening disease.

**SURGICAL CARE:** Excisional lymph node biopsy for the purpose of confirming the diagnosis is the only surgery indicated in Kikuchi's disease.

**CONCLUSION:** We describe a case of Kikuchi -Fujimoto disease presenting a unusual cause of posterior triangle cervical lymphadenopathy that mimicked tuberculosis lymphadenitis in a 16 years old female diagnosed by excision biopsy, treated with conservative treatment, because of its rarity and atypical presentation it is presented.

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