

# Kikuchi Disease : A Case Report

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## Introduction

Histiocytic necrotizing lymphadenitis (HNL), also known as Kikuchi disease is a rare condition of unknown etiology. Patients present with cervical lymph node enlargement, fever and malaise. The diagnosis is made by excision biopsy. However, this entity must be distinguished from both reactive processes and malignant tumors such as lymphoma. The clinical course is self limited with spontaneous resolution within a few months.

## Case report

This 18 years old female patient was admitted with complaints of fever and swelling on right side of neck since 20 days. She also gave history of profound weakness and bodyache. There was no history of cough, haemoptysis, rhinitis or sore throat. There was no history of jaundice. She gave history of loss of appetite. She had taken treatment from a private practitioner but did not get relief. FNAC from cervical lymph node was done and it reported inflammatory reactive lymphadenitis. She was empirically started on antitubercular treatment EHRZ + 4 HR. following which she developed severe vomiting.

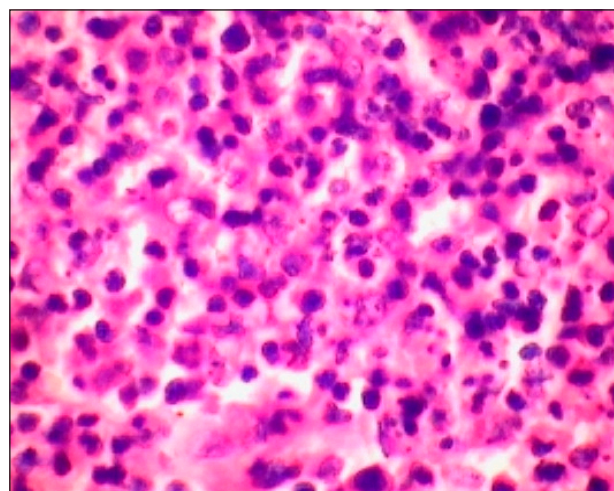
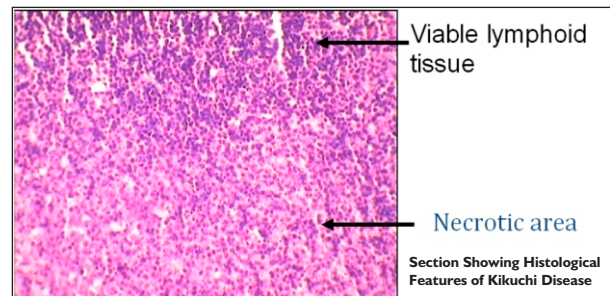
On examination she was febrile, toxic, had tachycardia, pallor but no icterus. She had palpable cervical lymph nodes on right side which were 3 to 4 in number, mobile, firm in consistency and tender (FIGURE 1). They appeared to be matted & overlying skin was normal. Throat examination was normal. There was no rash. Her systemic examination was normal. She was dehydrated.

On investigations her Haemoglobin was 8 gms / dl, TLC = 2,500 / cu mm, P= 72 %, L= 23%, E = 4% and M=1%. Peripheral smear was negative for malarial parasite and showed normocytic hypochromic RBCs ESR was 15 mm at 1 hour by wintrobe method. X-ray chest did not show any abnormality.

Excisional lymph node biopsy was performed. On gross examination lymph nodes were extremely fragile and could be removed in pieces only. However there was no



caseation seen. On microscopic examination of sections areas of necrosis and inflammation were seen with plenty of histiocytes (FIGURE 2 and 3). Hence a final diagnosis of Kikuchi's disease was made. Her ANA, RA factor was



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negative.

Patient was initially treated symptomatically. Her antitubercular drugs were stopped. She was also given 1 unit of Blood transfusion. After the report of biopsy she was given Prednisone 1 mg / kg in tapering doses for 3 weeks along with Diclofenac Sodium and Paracetamol tablets for pain and fever for first 3-4 days. Patients symptoms resolved and lymph nodes also decreased in size. She was completely alright after 3 weeks and is well and healthy till date.

### Discussion

Kikuchi-Fujimoto disease is a rare disease first described in 1972 by Kikuchi and Fujimoto et al. <sup>2</sup> However cases have been reported from all parts of the world. Clinically the disease presents with lymphadenitis usually in the cervical region.

Most reported cases of Kikuchi-Fujimoto disease have been of Asian origin. The cause is unknown and the condition is self-limiting. Some kind of viral or postviral etiology has been implicated. Bacterial and protozoal organisms as well as various other antigens, chemical, physical and neoplastic, have also been postulated. An association with systemic lupus erythematosus has also been shown <sup>5</sup>.

Kikuchi disease occurs in a wide age range of patients (ie, 2-75 y), but it typically affects young adults (mean age, 20-30 y). Women are affected more often than men by a ratio of 3:1.

A flu like prodrome with fever is present in 50 % of cases. Less commonly patient may have headache, nausea, vomiting, hepatic and pulmonary involvement, night sweats and rash may occur in 30%. Cervical lymphadenopathy is seen in majority of cases (80%). Posterior cervical nodes are frequently involved (65-70%). Lymphadenopathy is isolated to single location in majority of cases, but rarely multiple chains may be involved. Generalized lymphadenopathy is unusual. Lymph nodes are firm and non tender. Skin involvement in the form of maculopapular lesions, morbilliform rash, nodules, urticaria, and malar rash, which may resemble that of SLE may occur in some patients. Hepatosplenomegaly is rare. Neurological involvement may be seen in few cases in the form of aseptic meningitis or encephalitis <sup>3</sup>.

On investigation patients have leucopenia, raised ESR,

raised LDH levels. Diagnosis is usually done by histopathological examination of lymph nodes. It reveals a patchy necrosis with histiocytes and a fair amount of viable lymphatic tissue. Two histologic types are classically described, proliferative and necrotic types <sup>4</sup>. A study has also reported CT findings in Kikuchi's disease. CT features were perinodal involvement, homogenous contrast nodal involvement, local flow attenuation in lymph nodes and ring shaped lymph nodes <sup>7</sup>.

The course of Kikuchi disease is generally benign and self-limited. Lymphadenopathy most often resolves over several weeks to 6 months, although the disease occasionally persists longer. The disease recurs in about 3% of cases. Three deaths have been reported that occurred during the acute phase of generalized Kikuchi disease. One patient died from cardiac failure; the second, from the effects of hepatic and pulmonary involvement; and the last, from an acute lupus like syndrome. Another fatality has been reported in which Kikuchi disease appeared concurrently with SLE and was complicated with hemophagocytic syndrome and severe infection.

The treatment of Kikuchi's disease is basically supportive. However there is good response to short course of steroids like oral prednisone. Non steroidal Anti-inflammatory drugs are useful for controlling fever and pain <sup>6</sup>.

Our patient showed all the clinical features of Kikuchi disease and responded to steroids. This case is being reported to make the physicians aware of this entity. Kikuchi disease is underdiagnosed and hence underreported. Many a times patients may be put on antitubercular treatment thus exposing them to risk of drug induced hepatitis. This condition must be included in the differential diagnosis of cervical asymptomatic masses. The clinician must be aware of it to avoid long-term, costly treatments. Early recognition will minimize unnecessary investigations and prolonged empirical treatments.

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