

## Case Report

### Kikuchi's Disease: A Case Report

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#### Abstract :

Kikuchi's disease is also known as histiocytic necrotizing lymphadenitis. Kikuchi-Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis, is a rare benign, self-limiting cervical lymphadenitis of unknown etiology. It predominantly affects young women and can closely mimic infective and immunological disorders. Recognition of this condition is crucial, specially because it can easily be mistaken for tuberculosis, lymphoma, or even adenocarcinoma. We report an 18 years old girl who presented with fever and cervical lymphadenopathy. She had multiple enlarged cervical lymphadenopathy. Examination of other systems was normal. Laboratory investigations were also normal. Fine needle aspiration cytology of the cervical node was not done. The patient took a course of oral antibiotics. Since the patient did not respond, lymph node biopsy was done and the histological features suggested the diagnosis of Kikuchi's disease. The Patient was treated symptomatically and complete remission occurred in few weeks. Although the incidence of Kikuchi-Fujimoto disease is rare, clinicians should be aware of this condition as early recognition of the disease will minimize potentially harmful and unnecessary evaluations and treatments.

**Key words:** Kikuchi-Fujimoto disease, Histiocytic Necrotizing lymphadenitis, Systemic Lupus Erythematosus.

#### Introduction :

Kikuchi-Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis is an uncommon, idiopathic, generally self-limited cause of lymphadenitis<sup>1,2</sup>. Kikuchi first described the disease in 1972 in Japan. Fujimoto and colleagues independently described Kikuchi's disease in the same year. Kikuchi-Fujimoto disease is mainly seen in Japan. Isolated cases are reported in America, Europe and Asia. It is mainly a disease of young adults (mean age, 20-30 years), with a slight bias towards female<sup>3</sup>. Compared to male, ratio is of 1:4<sup>4</sup>. The cause of this disease is not known although infection and autoimmune etiologies have been proposed. Course of the disease is generally benign and self-limiting. Lymphadenopathy most often resolves over several weeks to 6 months. Recurrence rate is about 3%<sup>4</sup>. Mortality is extremely rare and usually due to hepatic, respiratory or cardiac failure. Awareness of this disorder will help to prevent misdiagnosis and inappropriate treatment.

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#### Case Report :

An 18 year old girl Mst. Jannati of Saltha, Faridpur district presented to us with multiple neck swellings and fever of 2 months duration. There was no weight loss. There was no previous history of tuberculosis or contact with tuberculosis. She did not have history of any drug intake or atopy. She did not have any other significant medical problems.

Clinical Examinations revealed bilateral large, mobile and tender-cervical lymphadenopathy, larger node being the right posterior triangle of neck which was measured about 2.5x4 cm, other enlarged nodes were right posterior cervical (2x2.2cm), left posterior auricular, left submandibular and right submandibular region. Lymph nodes were not palpable in other parts of the body. The blood pressure was 110/70 mm of Hg and the pulse rate was 80/min. Her cardiovascular, respiratory and neurological examinations were normal. The abdomen was soft with normal bowel sounds. Ear, nose and throat examinations were normal. Routine haematological parameters like hemoglobin, complete blood count, peripheral smear were within normal limits. ESR was 20mm in 1st hour. Blood glucose, urea, creatinine, sodium, potassium and bicarbonate levels were normal. Renal and liver function tests were normal. Blood and urine cultures were negative. Montoux test showed induration of 4mm. Ultrasound of abdomen and Chest radiograph was normal. Antinuclear antibody (ANA) and anti-ds DNA antibody were negative. Staining for AFB (acid-fast bacilli) was

also negative. Fine needle aspiration cytology (FNAC) of lymph node was not done. The patient was started an oral antibiotic (cap. cefixime-200mg 12 hourly for 7 days). Since the patient continued to have fever and persistent lymphadenopathy despite one week of antibiotic, lymph node biopsy was done and the histological features suggested the diagnosis of Kikuchi-Fujimoto disease (Figure 1). The patient was treated symptomatically with non-steroidal anti-inflammatory drugs, the lymph nodes regressed in one month.

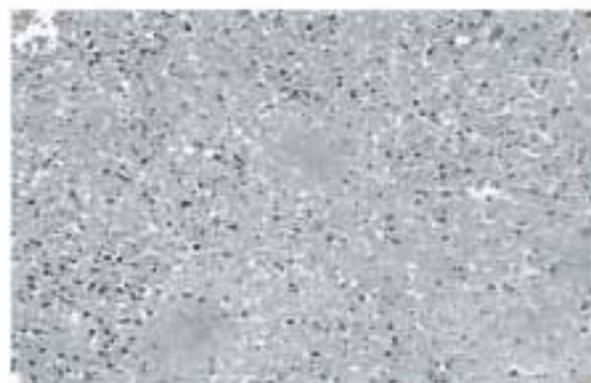


Figure 1: Lymph node biopsy section showing patchy areas of necrosis, proliferation of pale histiocytes, increase number of apoptotic cells, cellular debris and nuclear dust (karyorrhexis) (H&E, x100)

#### Discussion :

Kikuchi's disease most often presents with cervical lymphadenopathy which may be tender or non-tender and can be accompanied by fever, upper respiratory tract symptoms. Less common symptoms include arthralgia, skin rashes, weakness and night sweats. Weight loss, diarrhoea, anorexia, chills, nausea, vomiting, chest and abdominal pain have also been reported. Some patients may also have hepatosplenomegaly<sup>3-7</sup>. Involvement of skin and bone marrow has also been reported in some cases<sup>8,9</sup>. The exact aetiology of Kikuchi's disease is not known. Viral agents such as Epstein barr virus (EBV), Human immunodeficiency virus (HIV), Herpes simplex virus, dengue virus, Human T lymphotropic virus 1 (HTLV1) and Parvovirus B19 have been suggested as possible aetiological agents, but none have been confirmed so far<sup>4</sup>. Toxoplasma and other bacterial agents like *Yersinia enterocolitica*, *Bartonella*, *Brucella* have also been implemented<sup>9</sup>. An autoimmune mechanism has also been proposed because KFD is seen in conjunction with systemic lupus erythematosus (SLE). There are several reports suggesting an association between Kikuchi's disease and systemic

lupus erythematosus (SLE). However no convincing evidence is available to confirm such association. The pathogenesis of Kikuchi's disease is still not fully clear. It is supposed that the primary event may be the activation of T lymphocytes and histiocytes. Proliferating T cells enter the cycle of apoptosis, which may form the areas of necrosis in lymph nodes and then the cellular debris is removed by histiocytes<sup>10</sup>.

Routine laboratory investigations usually does not aid in the diagnosis of KFD except for erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) may be elevated in some cases and many patients have a low white blood count. Moreover, 25% to 31% of patients have atypical peripheral blood lymphocytes<sup>11</sup>. Fine-needle aspiration cytology (FNAC) only has a limited role in establishing the diagnosis of Kikuchi's disease. Diagnosis is based on histopathological findings of a lymph node biopsy.

Clinically Kikuchi's disease may mimic systemic lupus erythematosus (SLE) or Lymphoma (specially T-Cell non-Hodgkin's Lymphoma) as both this diseases can present with lymphadenopathy and fever and the skin lesions of Kikuchi's disease patients can resemble those seen in SLE. Careful histopathologic examination will thus help us distinguish KFD from other diseases. Histological feature which helps in the differentiation of KFD from the lymphadenopathy of systemic lupus erythematosus is almost total absence of plasma cells in the involved nodal tissue. Moreover appropriate serologic tests should be done to exclude systemic lupus erythematosus<sup>12</sup>. Features that distinguish KFD from malignant lymphoma include incomplete architectural effacement with patent sinuses, presence of numerous reactive histiocytes, relatively low mitotic rates, absence of Reed- Sterberg cell<sup>13</sup>.

The CT appearance of Kikuchi's disease may be variable, mimicking various nodal diseases, such as tuberculosis, metastasis and lymphoma. The necrotic area of the lymph nodes had a lower signal than the non-necrotic portion on T<sub>2</sub>-weighted MR images, which is different from the usual bright signal, intensity of nodal necrosis seen in other nodal diseases.

No specific treatment is available for Kikuchi's disease. Treatment is largely supportive. Non-steroidal anti-inflammatory drugs (NSAIDs) for tender lymph nodes and fever. The use of corticosteroids has been recommended in severe form of disease<sup>14</sup>. Intravenous immunoglobulin has also been tried with some success<sup>15</sup>. The disease usually runs a benign course and the condition is self-limiting, usually resolves in several weeks to months. The disease has a recurrence rate of 3% to 4%<sup>9</sup>.

**Conclusion :**

Although the incidence of Kikuchi's disease is rare, this disorder must be considered among the differential diagnosis when a young female patient presents with fever and cervical lymphadenopathy. Clinically Kikuchi's disease may mimic lymphoma or systemic lupus erythematosus (SLE). Therefore a careful histopathological examination is necessary in arriving at the diagnosis. Early recognition of the disease is very necessary for harmful and unnecessary evaluations and treatments.

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