



## KIKUCHI DISEASE WITH ASEPTIC MENINGITIS: A CASE REPORT

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**ABSTRACT**

**Introduction:** Kikuchi disease is a benign condition, usually affecting the lymph nodes. It mimics symptoms of other diseases and is commonly misdiagnosed.

**Case Report:** A 13 year old child presented with history of fever and right side cervical lymph node enlargement. Diagnostic procedures were performed to rule out infections, malaria and typhoid. Based on the presence of lymphadenitis and absence of any other disease, diagnosis of Kikuchi disease was made. After discharge, child presented within 4 days with positive meningeal signs. The final diagnosis was that of Kikuchi disease with aseptic meningitis.

**Conclusion:** The clinician needs to think of Kikuchi disease in the absence of a clear diagnosis and when there is presence of lymphadenitis. Counselling must be provided to patients diagnosed with this disease, to report if there are any new symptoms. Autoimmune disorders may precede or follow diagnosis.

**KEYWORDS :****INTRODUCTION**

Kikuchi disease is a rare lymphohistiocytic disorder, which is also known by the other names such as Kikuchi-Fujimoto disease(KFD) or necrotising lymphadenitis. This disease was first described by Fujimoto and Kikuchi in the year 1972. Kikuchi disease is a benign condition of the lymph nodes. It mimics tuberculous lymphadenitis and several other benign and malignant conditions. The disease is commonly misdiagnosed or undiagnosed, which leads to delays in the treatment. Kikuchi disease is self-limiting and the symptoms resolve within a few months, with or without treatment.

**EPIDEMIOLOGY:**

The incidence of Kikuchi disease is unknown mainly due to wrong diagnosis of the disease and also lack of diagnosis in several cases (1). Some studies suggest that young Asian females may be at a higher risk, but recent studies seem to indicate that adult men and women are affected equally and there is no difference in the ethnicity of those affected (1,2). A review of several literature suggests that in the paediatric population, males may be predominantly affected (3,4, 5).

**Aetiology:**

The exact cause of Kikuchi disease is unknown, however researchers have suggested that the cause of the disease can be viral infection, autoimmune or genetic.

**Case Study:**

The patient was a 13-year-old adolescent boy from an urban location who presented with a history of fever for 10 days. The child presented with fever that was low grade initially and then became high grade with chills and rigors. He had complaints of neck pain and swelling in the neck for 4 days. He also reported 6 episodes of non-projectile, non-bilious and non-blood stained vomiting.

On examination, the child was conscious, alert and not in distress. Temperature was 100.2 degrees Fahrenheit, Heart rate was 160 per minute, Respiratory rate was 22 per minute and BP was 106/60 mm of Hg. On systemic examination, child had 2-3 right cervical lymph nodes, that were 2.5cm x 1cm size, non-matted and tender. Bilateral tonsils were enlarged. There was no organomegaly and there were no neurological deficits.

**Diagnostic Procedures**

Tests were done to rule out malaria and typhoid. There was no growth in the blood culture. There was no focus of infection in the ultrasound scan of the abdomen. The TB work up was negative.

The blood test reports on the day of admission showed DC N54, L 34, M-8, platelets-1.5 lakhs, Sodium level was 139, Potassium was 4.6, Creatinine level was 0.74 and SGPT was 15. The WBC count was 3400.

Due to the history of prolonged fever and leukopenia, bone marrow was done which was a normal study. Lymph node biopsy was done. Sections showed lymph nodes with paracortical expansion and subcapsular wedge shaped necrosis with karyorrhectic nuclear debris surrounded by transformed lymphoid cells, histiocytes and plasmacytoid mononuclear cells focally forming aggregates. Special stains for acid fast bacilli and fungal organisms were negative. Necrotising lymphadenitis was seen in right cervical lymph node biopsy.

The diagnosis of Kikuchi disease was made based on the presence of lymphadenitis and symptoms consistent with Kikuchi disease. The child was started on Naproxen. The fever settled within 48 hours and child was discharged on naproxen. At discharge, adequate counselling was given to the child and parents to report if there were any abnormal changes in the health status.

**Sequel**

4 days post the initial discharge, the same child presented to Paediatric casualty with vomiting, fever and headache. On examination, meningeal signs were positive. The CT report showed no obvious abnormality in the brain parenchyma. There were no features suggesting raised intracranial tension. There was no ventricular dilatation or midline shift. CSF analysis was done and it showed the following counts – WBC:-200/cumm, RBC-160/cumm, CSF glucose was 45 and CSF protein was 112. There was no growth seen in the CSF Culture, Latex: Pneumococcal antigen detection in CSF- Negative.

The final diagnosis was that of Kikuchi disease with aseptic meningitis. The child was started on IV Antibiotics, Injection Cefotaxime 200mg/kg/day every 6 hours for 5 days and Injection Ceftriaxone 2gm IV twice a day for 5 days. Supportive treatment with Tab Naproxen 500 mg-, Tab Paracetamol 500 mg PRN and Injection Pantoprazole 40 mg OD was given. The condition of the child improved and was discharged with adequate counselling to report to the clinic if there was recurrence of symptoms or presence of any new symptoms.

**DISCUSSION:**

Kikuchi disease is a relatively rare disease. The disease mimics several other diseases and emergency physicians should be aware of this disease because it can be misdiagnosed commonly (6).

Patients presenting may have several other symptoms, but the disease is usually identified with the presence of lymphadenopathy. The lymphadenopathy is usually less than 3 cm, but there are instances of it being even 5 to 6 cm (7). Cervical lymphadenopathy is the most common occurrence, though axillary as well as supraclavicular lymphadenopathy have also been noted. Affected patients usually suffer from mild to moderate fever associated with headache, fatigue.

Many patients suffer from some skin conditions like occurrence of rashes, papules and plaques. Skin involvement in Kikuchi disease is usually indicative of severity of the disease (8).

The occurrence of aseptic meningitis in a patient affected by Kikuchi disease has been noted previously (9). There are also instances of recurrent aseptic meningitis associated with Kikuchi disease (10). In a retrospective literature review of 91 patients with review of literature, it was noted that there were only 2 cases of aseptic meningitis (11). So, it can be deciphered that the occurrence of aseptic meningitis in Kikuchi disease is a relatively rare presentation.

There may be spontaneous resolution of fever and lymphadenopathy in some of the patients, but in some, there can be recurrence of symptoms. Few of the patients diagnosed with Kikuchi disease may suffer from an underlying autoimmune disorder (12). The patient needs to be investigated thoroughly to rule out autoimmune disorder. Some patients may also develop such disorders after discharge. To ensure that this is identified and treated early, the patients need to be counselled to report to the clinic as soon as any symptoms appear or if there is any change in health status. Early diagnosis and long term follow up is necessary to reduce the risk of developing complications (13).

Treatment is usually based on the presenting symptoms. Since there is no specific guideline available for the treatment of this disease, physicians base their treatments on their experience with similar patients. Use of a combination of corticosteroids and hydroxychloroquine have been found to be effective in the treatment of Kikuchi Disease (14). Non-steroidal anti-inflammatory drugs are also used in the treatment of this disease (15).

#### CONCLUSION:

Kikuchi disease can be very distressing to the patient and the family. The presence of lingering fever, fatigue and other symptoms along with the delay in diagnosis can be very stressful. Since the disease mimics other diseases, the clinician should have this disease on mind if there is no other conclusive diagnosis, especially if the patient has lymphadenopathy (16). It is important to note that there are some patients diagnosed with Kikuchi disease with associated aseptic meningitis having a recurrence. This needs to be informed to all patients who are diagnosed with Kikuchi disease. Diagnostic tests to assess meningeal signs need to be included as a routine for all who are diagnosed with this disease.

Though studies have identified that young males may be predominantly affected than females in the paediatric population, further studies and analysis of data are recommended. In Asian communities, the male child is given priority and immediate healthcare is provided and the girl children are not given the same level of care, which may lead to underreporting of cases.

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