

## Case Report

# Kikuchi disease – a rare cause of pyrexia of unknown origin

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## Introduction

Kikuchi-Fujimoto disease or Kikuchi disease is a disease of unknown aetiology with a benign self-limiting course presenting with tender lymphadenopathy with or without systemic manifestations including low grade fever. Recurrence of disease is rare, and it rarely causes fatalities [1]. We report a case of Kikuchi disease presenting as pyrexia of unknown origin.

## Case report

A 21-year-old girl presented with a history of fever of one-month's duration associated with malaise, reduced appetite and generalized body aches. She did not have dysuria, abdominal pain, respiratory symptoms, headache, or altered bowel habits. She denied any photosensitive rash, oral ulcers, hair loss or any other features suggestive of autoimmune disease. Her past medical history was insignificant and she denied any recent travel history or exposure to tuberculosis. On examination, her temperature was 102°F, pulse rate was 120bpm and blood pressure was 120/80mmHg. Palpable, tender, enlarged posterior cervical lymph nodes were noted bilaterally. Cardiovascular and respiratory examinations were unremarkable and there was no palpable organomegaly on abdominal examination.

Full blood count revealed a white blood cell count of  $2.44 \times 10^3/\mu\text{L}$  (neutrophils–52.2%, lymphocytes–38.1%, eosinophils–2.2%), haemoglobin of 10.0g/dL and platelet count of  $334 \times 10^3/\mu\text{L}$ . ESR was 53mm/1<sup>st</sup> hour and CRP was 4.3mg/L. Renal functions and liver functions were within normal limits. Peripheral blood film showed bicytopenia with moderate rouleaux formation, suggestive of an infective or inflammatory cause with evidence of possible iron deficiency anemia. Lactate dehydrogenase (LDH) level was 395.2U/L (225-450), ferritin was 259ng/mL (5-148) and thyroid functions were within normal limits. Antinuclear antibody, dsDNA antibody and rheumatoid factor were negative. Blood culture revealed no growth. Echocardiography did not reveal any valvular defect or vegetation. Three samples of sputum for acid fast bacilli (AFB), chest radiography and the Mantoux test were negative for tuberculosis. Serological studies for Epstein Barr virus, cytomegalovirus and toxoplasma did not show any active infection.

Trephine biopsy of bone marrow was reported as 'reactive bone marrow secondary to a chronic infective or inflammatory process'. Bone marrow aspiration was negative for fungi by direct microscopy and culture. Lymph node biopsy of a cervical lymph node showed extensive areas of necrosis associated with abundant nuclear debris. It also showed a collection of histiocytes in the vicinity of necrosis. The histology was compatible with necrotizing lymphadenitis. A diagnosis of Kikuchi disease was made after exclusion of systemic lupus erythematosus (SLE) and lymphoma. Patient was started on naproxen 250mg eight hourly for two weeks and the fever subsided within a couple of days. Patient improved clinically and did not develop fever again.

## Discussion

Kikuchi disease was identified in 1972, in Japan, by the pathologists Kikuchi and Fujimoto, independently. It mainly affects young Asian adults with a female predominance [2]. It is a benign mimic of serious conditions such as systemic lupus erythematosus, lymphoma and tuberculous lymphadenitis which need careful exclusion prior to diagnosis as a Kikuchi disease [1,2,3]. Since there are no immunohistochemical markers for Kikuchi disease available at present, diagnosis is made by excluding other serious conditions. It typically presents with cervical lymphadenopathy and low-grade fever. Fever is a main symptom in 30-50% of cases and is low grade in nature, lasting up to one week [4]. Our patient presented with a high-grade fever lasting more than one-month. Cervical lymph nodes, especially posterior cervical lymph nodes, are commonly involved with Kikuchi disease and generalized lymphadenopathy is rare.

Laboratory and radiological investigations are nonspecific and excision biopsy of a lymph node assists in confirmation. Typical histological features to suggest Kikuchi disease are patchy or confluent paracortical necrosis, presence of histiocytes, partial effacement of the nodal architecture, absence of neutrophils, plasmacytoid monocytes, presence of karyorrhectic debris, and eosinophilic apoptotic debris. Absence of granulocytes and granuloma also helps to differentiate from other conditions [1]. Inflammatory markers such as ESR and CRP can be elevated and granulocytopenia can occur in Kikuchi disease. [1] ANA and rheumatoid factor are usually negative.

Treatment is generally supportive and giving nonsteroidal anti-inflammatory drugs (NSAIDs) helps to alleviate nodal tenderness and fever. We have successfully managed our patient with naproxen. Corticosteroids such as prednisolone can be used in severe or extra nodal cases [5]. Steroid resistant cases are treated with intravenous immunoglobulin and hydroxychloroquine [1].

## Conclusion

Although Kikuchi disease is a rare condition, it should be considered as a differential diagnosis in a young patient coming with fever and cervical lymphadenopathy. However, the patient should be reviewed in two to three months, to ensure cessation of symptoms, since, very rarely, it can evolve into a lymphoma.

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### **Declaration of conflicting interests**

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

### **Informed consent**

Informed written consent was obtained from the patient for her anonymized information to be published in this article.

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