

# A rare case of Kikuchi-Fujimoto disease (case report)

## Case Report

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## Abstract

**Background:** Kikuchi disease is a rare, benign, self-limited disease characterized primarily by fever and cervical lymphadenopathy. Diagnosis is based on excisional biopsy and pathologic study. We report a case of an atypical axillary lymph node enlargement.

**Case report:** This patient was a 12-year-old boy with clinical characteristics including axillary lymph adenopathy, fever and fatigue. He became asymptomatic after excisional biopsy. Histologic study reported necrotizing lymphadenitis without neutrophils (Kikuchi disease).

**Conclusions:** These findings are important for diagnosis, because of the disease rarity, clinical features (such as lymphadenopathy, prolonged fever) and unidentified etiology.

**Key Words:** Lymphadenopathy, Fever, Kikuchi-Fujimoto Disease, Childhood.

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## Introduction

Kikuchi disease also known as histiocytic necrotizing lymphadenitis or kikuchi Fujimoto disease (KFD) is a benign and self-limited disease, which has been first reported in Japan in 1972 [1,2,3]. It is rare and has a worldwide distribution with a higher prevalence among Japanese and other Asiatic individuals [4]. Pileri et al. reported 23 cases from Germany, Spain, Italy, Iran, and South Korea [5]. The initial presentation includes cervical lymphadenopathy, fever, skin rash, headache, fatigue, leukopenia [6], rarely night sweat, weight loss, aseptic meningitis, myocarditis, pneumonitis, hepatosplenomegaly and acute kidney injury [7, 8, 9], and more often, it occurs in children 8-16 yr of age [3]. Its etiology has remained unknown [3], but infectious agents including Epstein – Barr virus (EBV), cytomegalovirus, human herpes virus 6 and 8, parvovirus B19, Toxoplasma, Yersinia have been reported to have a causative role, but no specific agents have been identified [10, 11]. KFD is a self-limited benign condition that spontaneously appears to resolve within weeks to 6 months; however, 3-4% of patients will experience recurrent episode of KFD. In spite of its rarity in the pediatric population, KFD clinically should be differentiated from other infectious diseases (such as cat scratch disease, bacterial lymphadenitis and tuberculosis), lymphoma and systemic lupus erythematosus (SLE). The results of a wide range of laboratory tests are usually normal in patient with KFD. Anemia, mild leucopenia and slight elevation of ESR are the most common abnormal laboratory findings [12]. Diagnosis of KFD is based on excisional biopsy and histological examination [5-7]. The pathological features of KFD include lymph node necrosis with karyorrhexis surrounded by histocytes without granuloma neutrophil or plasma cell infiltration [5]. Immunohistologic study of lymph node may reveal the large cells, in KFD are CD8-positive immunoblasts and CD68-positive histiocytes [7]. Treatment is suggested unnecessary and not recommended unless there is coincidence with SLE [6].

In this study, we report a case of an atypical axillary lymph node enlargement in an Iranian child.

## Case Report

A 12-year-old boy was admitted to our center because of a 5-day history of fever, fatigue and left axillary lymph node enlargement 2 weeks before admission. The patient had no complaints of sore throat, night sweat, and headache.

Clinical history did not reveal any travel, exposure to animals or insect bites. On admission, the patient was in good general condition even with an axillary temperature of 40°C. On examination, there was only unilateral, non-fluctuant swelling, immobile and tender 1\*1.5 cm lymphadenopathy and no skin rash on the trunk and limbs. In addition, there was no hepatosplenomegaly or palpable mass in abdominal exam.

Other physical examinations were normal.

Complete blood cell demonstrated a white blood cell count of  $3.3 \times 10^9/L$ , with 43% neutrophil, 54% lymphocyte, 2% monocyte, 1% eosinophil and platelet count of  $64 \times 10^9/L$  and hemoglobin was 11.4 mg/dl. Urine analysis, liver and renal function tests, ESR and CRP were normal and blood culture was negative for bacterial and fungal infection. The results for EBV capsid antigen, PPD test, Wright, Coombs Wright and Widal tests, ANA, anti-ds-DNA and chest x-ray and abdominopelvic sonography were normal.

Bone marrow aspiration studies were normal without any abnormal cells or excess blasts. Intravenous antibiotics were initiated immediately. Fever continued 13 days after hospitalization and using intravenous antibiotics; therefore, the excisional biopsy was done. Fever stopped 24 hours after excisional biopsy.

Histologic study reported necrotizing lymphadenitis without neutrophils (Kikuchi disease).

Immunohistologic study was reported that CD3 polyclonal rabbit anti-human was strongly positive, CD20 (L26) was weakly positive, CD68 clone and PG-M1 in many cells were positive, CD30- Ki-1 antigen close ber H-Z was positive in non-atypical predominantly cell, but CD 10 clone and 56 C6 were negative.

## Discussion:

This study reported the clinical presentation of KFD by axillary lymphadenopathy, which was not

common. Although KFD has shown a female predominance, most pediatric studies have reported a male predominance [13-18]. Patients may present with fever, chills, weight loss, arthralgia, splenomegaly, abdominal pain, fatigue and skin rash [7, 12, 19-23]. Our case was a 12-year-old boy in good condition with fever, fatigue and unilateral axillary lymph node about 2 weeks before admission.

However, the most common presenting symptom, unilateral cervical lymphadenopathy, has been reported [12, 22-27]. In only a few cases, the lymphadenopathy involved the peritoneal and supraclavicular nodes [12, 20, 21]. The affected node in our patient was unilateral non-fluctuant, swelling immobile and tender in axillary.

The size of lymph nodes was usually less than 3 cm in previous reports [11, 12, 24-26]; however, they may reach 5-6 cm in diameter [7]. The size of lymph node in our patient was 1\*1.5 cm.

Approximately 40% of patients experienced leukopenia ( $<4000/\mu l$ ) and 15% presented with thrombocytopenia ( $<150000/\mu l$ ) based on Kim's report [26]. Our patient had leukopenia ( $3300/\mu l$ ) and thrombocytopenia ( $64000/\mu l$ ) which were not common in the previous study [24, 26].

Unlike the study of Dumas et al., the reported patient in the current study was male and his disease was not severe [27].

Diagnosis of KFD was based on excisional biopsy and histological study in all cases [5-7, 12, 19, 22, 24-27].

Immunohistochemistry staining study of lymph node for KFD indicated that CD4, CD8, CD3 were positive but CD11-b was negative [28]. In another study, CD20, CD 10, CD3, CD43, and CD30 were positive [6]. Immunohistochemistry staining study from excisional lymph node biopsy in our case was positive in CD3, CD20, CD 68 and negative for CD10. Our patient became afebrile 24 hours after excisional biopsy and did not have other problems.

In conclusion, persistent lymph node enlargement with a prolonged fever requires a careful differential diagnosis which should include the possibility of KFD. Through careful diagnosis patients with KFD may avoid unnecessary treatment with aggressive chemotherapy. More research and attention should be directed towards the etiology of the disease.

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