

Kikuchi-Fujimoto Disease in 21-Year-Old Man

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ABSTRACT

Kikuchi-Fujimoto disease also known as histiocytic necrotizing lymphadenitis is a benign disorder characterized histologically by necrotic foci surrounded by histiocytic aggregates, and with the absence of neutrophils. The patient was a 21-year-old man with chills, fever and cervical lymphadenopathy. He had swelling and tenderness of cervical area. Multiple enlarged lymph nodes were palpable. Cervical lymph node excisional biopsy was performed. Microscopic examination showed reactive follicular hyperplasia with vast areas of necrosis without neutrophilic infiltration. No sign of malignancy was seen. Final diagnosis was Kikuchi-Fujimoto necrotizing lymphadenitis. Kikuchi-Fujimoto disease should be considered as one of the differential diagnoses in patients with prolonged fever and cervical lymphadenopathy. It should be differentiated from tuberculous lymphadenitis in regions where tuberculosis is prevalent.

Keywords: Cervical, Kikuchi, lymphadenitis

INTRODUCTION

The Kikuchi-Fujimoto disease, also known as histiocytic necrotizing lymphadenitis, is a self-limiting lesion of unknown cause first described in 1972 independently by Kikuchi and Fujimoto *et al.* This self-limiting disorder usually occurs in Asian women in their late 20 s or early 30 s. Typically, it runs a benign course and appears to resolve spontaneously 1 to 6 months after definite diagnosis. The Kikuchi-Fujimoto disease usually manifests as a localized cervical lymphadenopathy.^[1]

CASE REPORT

The patient was a 21-year-old man with chief complaints of chills, fever, cervical lymphadenopathy and myalgia. On admission vital signs were as: T = 39°C, PR = 90/min, RR = 15/min, BP = 100/60 mmHg. The patient was conscious and orient. Physical examination of chest, heart, abdomen, extremities and neurologic examination were normal. He had swelling and tenderness of cervical area and multiple enlarged lymph nodes were palpable. Examination of pharynx revealed no erythema or exudation. Tonsils were normal. Laboratory findings were as follow:

CBC: WBC=15700, Hb=13.6g/dl, Plt=209,000, ESR = 92 mm, CRP = 24 mg/l, BUN = 18 mg/dl, Cr = 1 mg/dl, Na = 143 meq/dl, K = 4.5 meq/dl, FBS = 78 mg/dl. ALT = 25 U/L, AST = 33 U/L, PT=12.5 s, PTT=28 s, ALP=278 U/L, LDH=1398 U/L, CPK = 52 IU/L, INR = 1. Urine analysis and peripheral blood smear were normal. Wright, widal, HBs-Ag and HIV-Ab were negative. EBV, CMV and toxoplasma antibodies (IgM and IgG) were negative.

Cervical soft tissue sonography showed multiple lymph nodes with different size. The largest was 9 * 15 mm.

Ceftriaxone and azithromycin was started for him. Cervical lymph node excisional biopsy was performed. Microscopic examination showed reactive follicular hyperplasia and some areas of monocytoid lymphoid cells hyperplasia with vast areas of necrosis without neutrophilic infiltration. No sign of malignancy was seen. Final diagnosis was Kikuchi-Fujimoto necrotizing lymphadenitis. Prednisolone 25 mg daily was started for him. Fever disappeared and general condition improved gradually. The patient was discharged with good condition and outpatient follow-up.

DISCUSSION

Kikuchi-Fujimoto disease is characterized by fever and lymphadenopathy, usually localized in the cervical region. This disease principally affects young females.^[2] It is also known as histiocytic necrotizing lymphadenitis, is a benign disorder characterized histologically by necrotic foci surrounded by histiocytic aggregates, and with the absence of neutrophils.^[3] 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. Lymphadenitis, hepatomegaly and splenomegaly, as well as leukopenia, elevated erythrocyte sedimentation rate and hepatic abnormalities are common findings. Fever, malaise, fatigue, headache, night sweats, nausea, vomiting, weight loss, cutaneous manifestations, and even neurological symptoms are other complaints.^[4] Diagnosis is fundamentally based on the affected

lymph node biopsy, since laboratory data are often normal.^[5] Final diagnosis could only be established on the basis of typical morphological changes in the lymph node, and lymph node biopsy is needed for establishing the diagnosis. Lymphadenopathy in a patient with fever of the unknown origin could provide a clue to the diagnosis of lymphoma, tuberculosis, metastatic carcinoma, toxoplasmosis and infectious mononucleosis.^[3] Histologically, the lesions affect the cortical and paracortical areas of the node. Characteristic features include focal necrosis predominantly in the paracortical region with abundant karyorrhectic debris and atypical mononuclear cells around the necrotic zone (crescent-shaped histiocytes, plasmacytoid monocytes, and small lymphocytes and immunoblasts, mostly CD3 (+)/CD8 (+)), most often with an intact lymph node capsule, an absence of neutrophils, and a paucity of plasma cells. Kikuchi-Fujimoto disease has been classified into three histological subtypes and is thought to progress from proliferative type (>50%) to necrotizing type (30%) and finally resolve into xanthomatous type (<20%). Differential diagnoses should include malignant lymphoma, infectious diseases such as toxoplasmic lymphadenitis, tuberculous lymphadenitis and cat scratch disease, and systemic lupus erythematosus (SLE).^[6] It has a benign evolution process, with a spontaneous healing in some weeks or months.^[5] Treatment is symptomatic (analgesics-antipyretics, non-steroidal anti-inflammatory drugs and, rarely, corticosteroids). Spontaneous recovery occurs in 1 to 4 months. Patients with Kikuchi-Fujimoto disease should be followed-up for several years to survey the possibility of the development of systemic lupus erythematosus.^[7] Kikuchi-Fujimoto disease should be considered as one of the differential diagnoses in patients with prolonged fever and cervical lymphadenopathy and that it should be differentiated from tuberculous lymphadenitis in regions where tuberculosis is prevalent.^[8]

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