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A Case of Recurrent Cervical Lymphadenopathy Due to Kikuchi-Fujimoto Disease

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Conflict of interest: None declared

Patient: Female, 52-year-old
Final Diagnosis: Kikuchi-Fujimoto disease
Symptoms: Cervical lymphadenopathy • fever
Medication: —
Clinical Procedure: —
Specialty: Infectious Diseases • Rheumatology

Objective: Unusual clinical course

Background: Kikuchi-Fujimoto (KFD) disease is a rare benign syndrome of necrotizing lymphadenopathy with distinctive histologic characteristics and was first reported in 1972. It is usually manifested by painful cervical lymphadenopathy and fever in young people, predominantly in females. KFD is a self-limiting disease and recurrence is rare.

Case Report: We present the case of a middle-aged woman who presented with recurrent cervical lymphadenopathy 3 times in 13 years. Diagnosis of KFD was made by excisional lymph node biopsy, showing typical histopathologic features. She was treated with oral prednisone, with a favorable response.

Conclusions: In patients with recurrent lymphadenopathy, excisional biopsy can lead to definitive diagnosis and directed treatment. KFD is an uncommon and potential cause for recurrent lymphadenopathy.

Keywords: Histiocytic Necrotizing Lymphadenitis • Histiocytosis, Sinus • Image-Guided Biopsy

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Background

Kikuchi-Fujimoto Disease (KFD) was first reported by Kikuchi and Fujimoto in Japan in 1972. It is also known as histiocytic necrotizing lymphadenitis. Clinical features are fever and painful cervical lymphadenopathy. It is seen more commonly in females [1,2]. Diagnosis of KFD requires biopsy and histology of lymph nodes [3]. Typical histological features of KFD include a focal proliferation of histiocytic cells and abundant nuclear debris and absence of neutrophils [4]. Most cases resolve within 1-3 months and recurrence is rare [5]. We present a case of undiagnosed recurrent cervical lymphadenopathy occurring 3 times in 13 years, which was found to be due to KFD.

Case Report

A 52-year-old Indian woman presented with a 7-day history of fever and painful right-sided cervical lymphadenopathy. The review of symptoms was otherwise unremarkable. The patient reported 2 similar episodes of self-limiting cervical lymphadenopathy and fever, occurring 7 and 13 years ago. She did not seek medical care on either occasion.

The patient was febrile (38.5°C) and other vital signs were within normal limits. Results of a physical examination were notable only for right-sided tender and firm cervical lymphadenopathy.

A complete blood count showed white blood cell count 7200/cmm, hemoglobin 12.6 g/dl, and platelets 140 000/cmm. She was found to have BUN 5.3 mmol/l, serum creatinine 60 umol/l, serum ferritin 500 ug/l, and C-reactive protein (CRP) 60 mg/l. Nasopharyngeal SARS-COV2 PCR was negative. Epstein-Barr viral antibodies, HIV, rheumatoid factor, and antinuclear antibodies (ANA) were negative.

A blood culture was sent and broad-spectrum antibiotics were started empirically. A neck ultrasound revealed multiple cervical lymph nodes; the largest was 1.9×0.9 cm on the right side. CT neck and chest showed right-sided multiple cervical lymph nodes; the largest was 2.8×2.3 cm (**Figure 1**).

The patient continued to be febrile despite treatment with broad-spectrum antibiotics. On day 3 of hospitalization, she underwent right cervical lymph node excisional biopsy, which showed nuclear debris and necrosis with infiltration of many histiocytic cells, and no neutrophils were seen (**Figure 2**). Lymph node bacterial, fungal, and tuberculosis cultures were negative. Based on pathology results, a diagnosis of KFD was established.

Broad-spectrum antibiotics were stopped and oral naproxen was initiated. Due to lack of response to oral naproxen for 3 days, we switched it to oral prednisone. After 3 days of oral prednisone, her fever subsided, with near complete resolution of right cervical lymphadenopathy. She was discharged home on a short course of oral prednisone. Outpatient follow-up assessment 1 week after discharge revealed complete resolution of cervical lymphadenopathy.

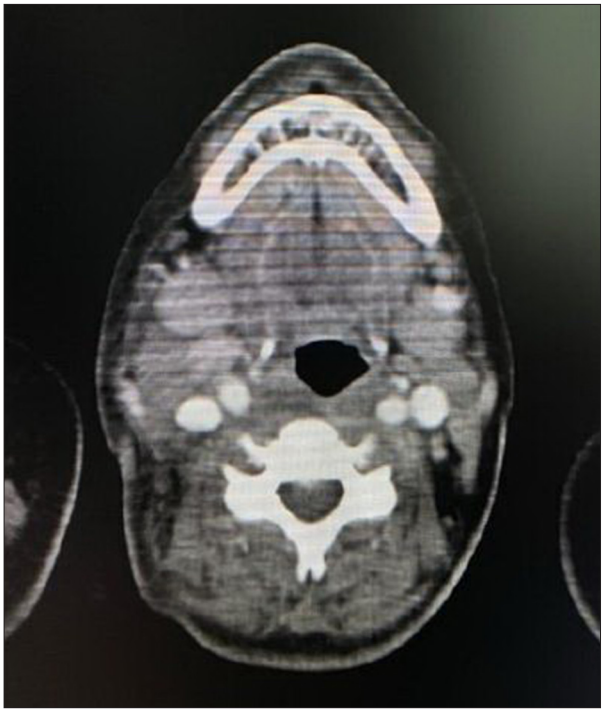

Figure 1. Axial section of CT neck showed right-sided multiple cervical lymph nodes; the largest was 2.8×2.3 cm.

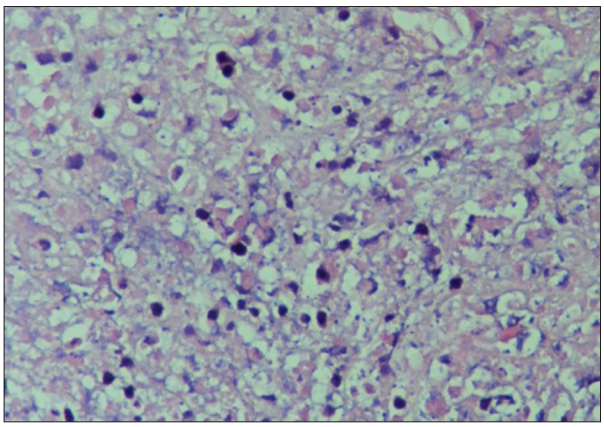

Figure 2. Histopathology of cervical lymph node showed nuclear debris and necrosis with infiltration of many histiocytic cells.

Discussion

KFD is a rare idiopathic cause of lymphadenopathy, first reported in 1972. The disease commonly presents with unilateral

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cervical lymphadenopathy in young people [4]. The size of lymph nodes is usually 1-4 cm, but up to 6 cm has been reported [6,7]. In our case, the size of the swollen lymph node was 2.8 cm.

Excisional lymph node biopsy is the preferred diagnostic method, as the disease may affect the lymph node partially. Typical histopathological features can distinguish KFD from other diseases. These features were seen in our case and confirmed the diagnosis.

Treatment of KFD is generally supportive and patients usually respond to non-steroidal anti-inflammatory drugs (NSAID). Similar to our case, corticosteroids have induced good response in patients who did not improve with NSAID [8,9].

Recurrence of disease seems to be rare and is reported in no more than 2-3% of patients. Our patient presented with recurrent symptoms 3 times over a period of 13 years. Her first and second episodes spontaneously resolved, but the current episode needed corticosteroid therapy to induce remission.

Conclusions

In recurrent febrile unilateral lymphadenopathy, excisional biopsy can lead to a definitive diagnosis and directed treatment. Recurrence of KFD is rare. Therefore, long-term follow-up of patients with KFD is needed.

Conflict of Interest

None.

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