CASE REPORT

KIKUCHI-FUJIMOTO DISEASE IN A 24-YEARS OLD SAUDI FEMALE IN SAUDI ARABIA

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ABSTRACT

Kikuchi-Fujimoto disease (KFD) is a very rare, self-limited and benign inflammatory disorder. It was first reported in Japan in 1972. Diagnosis of Kikuchi disease is confirmed by histopathological investigation of lymph node (LN) biopsy under ultrasound guidance. A 24-year old medically free female presented to the oncology out-patient clinic at King Abdulaziz University Hospital, Jeddah, Saudi Arabia complaining of a 7-days history of small, painless and slowly-growing, lift-sided neck mass. An excisional biopsy was taken from the mass and sent for histopathological analysis which confirmed the diagnosis of KFD. KFD patient need a regular follow up for the recurrence of the disease and to detect other autoimmune disorders that could be associated with KFD such as SLE. We describe a case of KFD affecting the cervical lymph nodes of a 24-year old Saudi female.

KEY WORDS: Kikuchi-Fujimoto disease, lymphadenopathy, autoimmune, head and neck surgery, head and neck oncology.

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INTRODUCTION

Kikuchi-Fujimoto disease (KFD), also known as histiocytic necrotizing lymphadenitis is a very rare, self-limited and benign inflammatory disorder. It was first reported in Japan in 1972 by Kikuchi and Fujimoto, Japanese researchers and physicians (1-2). KFD is an autoimmune inflammatory disorder that commonly involves the cervical lymph nodes with a high probability of recurrence (3-4). Most patients with KFD complain of pain and tenderness in the cervical lymph nodes, usually unilaterally. KFD can affect the axillary and inguinal lymph nodes as well but in a very low percentage. It can also manifest and present with a wide range of symptoms, from mild systemic symptoms to serious constitutional symptoms. Some of the serious symptoms that KFD can present with are high-grade fever, arthralgia and hemophagocytic lymphohistiocytosis (5-7). However, the etiological factors of KFD stay obscure and its presentation of lymphadenopathy and necrotic lesions usually lead to the misdiagnosis of KFD with other lymph nodes disorders such as head and neck infections, malignancies, metastasizes or lymphomas (3-6).

Diagnosis of Kikuchi disease is confirmed by histopathological investigation of lymph node (LN) biopsy under ultrasound guidance or by excisional biopsy. Previous literature has revealed that computed tomography (CT) or magnetic resonance imaging (MRI) findings help in diagnosis of Kikuchi cervical lymphadenopathy but don’t provide a definitive diagnosis for this condition (8-9).

KFD usually respond to medical therapy and that is mainly depends on the presentation and the associated clinical manifestations. Management of KFD include using non-steroidal anti-inflammatory and corticosteroids therapy to control the inflammation.

We are reporting in this manuscript a case of KFD affecting the cervical lymph nodes of a 24-year old female.
CASE REPORT
A 24-year old medically free female presented to the oncology out-patient clinic at King Abdulaziz University Hospital, Jeddah, Saudi Arabia complaining of a 7-days history of small, painless and slowly-growing, lift-sided neck mass. She had a history of fatigue, weight loss, undocumented fever for 2 weeks and shoulder pain. The patient denied having any shortness of breath, dysphagia, hoarseness or other respiratory or upper airways symptoms. She denied any joint pain or skin changes. She also denied any history of trauma, night sweats or temperature intolerance. She denied history of using. The denied any family history of head and neck cancer. She lives in Jazan in south of Saudi Arabia and she denied any type drug abusing or tobacco smoking. The patient denied surgical history and she was medically free. Physical examination revealed a solitary, non-tender, lift-side neck mass at level I without an overlying skin changes.

An excisional biopsy was taken from the mass and sent for histopathological analysis. Microscopic examination showed wide areas of necrosis, some of them are paracortical (Figure 1).

Figure 1: Sections revealed wide areas of necrosis. The center of necrosis contains abundant karyorrhectic debris and fibrin deposits surrounded by foamy histiocytes with crescent-shaped nuclei, atypical plasmacytoid monocytes and karyorrhexis.

The center of necrosis contained abundant karyorrhectic debris and fibrin deposits surrounded by foamy histiocytes with crescent-shaped nuclei, atypical plasmacytoid monocytes and karyorrhexis (Figure 2).

Figure 2: Coronal CT scan of the neck showing enlarged bilateral jugulo-digastric lymph nodes with evidence of necrotic changes in the left side.

Figure 3: Axial CT scan of the neck showing enlarged bilateral jugulo-digastric lymph nodes with evidence of necrotic changes in the left side.

A panel of immunohistochemical markers were performed including: CD3, CD20, CD68 and CD15. The atypical cells lack of monoclonal lymphocyte receptors that rules out the possibility of lymphoma. The cells took the stain of CD3, CD15 and CD68 that are consistent with plasmacytoid monocytes.

Computerized tomography (CT) scan for neck, chest and abdomen was done. CT scan of the neck demonstrated an enlarged bilateral jugulo-digastric lymph nodes in the left side with evidence of necrotic changes in the left side only (Figure 3,4).

Figure 3: Coronal CT scan of the neck showing enlarged bilateral jugulo-digastric lymph nodes with evidence of necrotic changes in the left side.

Figure 4: Axial CT scan of the neck showing enlarged bilateral jugulo-digastric lymph nodes with evidence of necrotic changes in the left side.

Otherwise there were no significant changes in the other lymph nodes or surrounding structures. Other investigations such as complete blood count (CBC) showed a microscopic hypochromic anemia with a hemoglobin level of 9.8 g/dL and low neutrophil count (1720 /mm3). The inflammatory parameters showed high C-reactive protein (CRP) (CRP=3.44 mg/L) but other investigations were normal. All the previous findings were consistent with the diagnosis of Kikuchi-Fujimoto disease.

Decision was made to start the patient on 5 mg prednisolone for five days and follow her up in 3 months. After 3 months the patient showed significant improvement. However, after 5 months the patient had recurrence and the CT scan of the neck and soft tissue showed no change in size of lymph nodes from the first CT scan and the decision was made to treat the patient the same as the first presentation and follow her up. A written consent and the approval of patient was taken before writing this report as well as an ethical approval from the unit of biomedical ethics at King Abdulaziz University Hospital.

DISCUSSION
KFD has a high prevalence among young ladies in Japan and other Asian nations. However, recent studies showed that male to female ratio of the disease are close to 1:1. (11) This may be related to the association of some genetic mutations with KFD which were found more in that population more than other areas. Recent literature showed that these genetic mutations might be related to mutations in human leukocyte antigen (HLA) class II genes (3,12).
Some researchers classify KFD as an autoimmune disorder because of the histopathological features of the disease. Previous literature reported the association of the disease with systemic lupus erythematosus (SLE). (3,4,10) Other studies have shown that the disease is associated with some infectious agents. However, there is still no clear evidence of the association of KFD with specific infections or other autoimmune disorders like SLE (20).

The differential diagnosis also includes malignancies such as lymphoma and other head and neck malignancies. However, in our case lymphoma was ruled out by histopathological studies. Other differential diagnosis include viral infections such as herpes simplex virus infections and infectious mononucleosis that is caused by Epstein-Barr virus which usually affect immunocompromised patients. Other infections include Toxoplasma gondii, necrotizing granulomatous lymphadenitis of tuberculosis, histoplasmosis, leprosy and cat-scratch disease (13). All of the above mentioned differential diagnosis were rolled out in our case by medical history, laboratory investigations and histopathological pattern of the disease. Most patients with KFD complain of unilateral cervical lymphadenopathy with a wide range of symptoms, from mild systemic symptoms to serious constitutional symptoms (high grade fever, arthralgia, and hemophagocytic lymphohistiocytosis) (5,7). In 2013, a case has been reported in UK of a patient who presented with a lump in the groin and was initially diagnosed with lymphoma and started treatment. However, after treatment the patient came back with the same presentation and finally diagnosed with KFD. (18).

**AUTHORS’ CONTRIBUTIONS**

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript and provided approval for this final revised version.

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