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Kikuchi-Fujimoto Disease Presenting with Supraclavicular Lymphadenopathy: A Case Report

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SUMMARY

A 34-year-old female who had supraclavicular lymphadenopathy applied to our clinic. Pathological examination of the lymph node excised via biopsy was reported as Kikuchi-Fujimoto disease (KFD) also called histiocytic necrotising lymphadenitis. The clinical course was self limited with lymph node excision and nonsteroid anti-inflammatory drugs. Spontaneous resolution was seen within two months as reported in the literature. KFD's etiology is unknown and it is one of the reasons of benign lymphadenopathy. It is important because of the possibility of misdiagnosis with many diseases including lymphoma.

Key words: *Kikuchi-Fujimoto, Histiocytic Necrotising Lymphadenitis, Lymphadenopathy*

ÖZET

Supraklavikuler Lenfadenopati ile Saptanan Kikuchi-Fujimoto Hastalığı: Olgu Sunumu

Supraklavikuler lenfadenopatisi olan 34 yaşında kadın hasta kliniğimize başvurdu. Biopsi ile çıkarılan lenf nodunun patolojik inceleme sonucu aynı zamanda histiositik nekrotizan lenfadenit de denilen Kikuchi-Fujimoto hastalığı (KFH) olarak raporlandı. Klinik gidiş lenf nodunun eksizyonu ve nonsteroid anti-inflamatuar ilaçlarla kendini sınırladı. Literatürde bildirildiği gibi iki ay içinde spontan iyileşme görüldü. KFH'nın sebebi bilinmemektedir ve benign lenfadenopati nedenlerinden biridir. Lenfoma dahil olmak üzere lenfadenopatiye sebep olan diğer pek çok hastalıkla karışabilmesi açısından önemlidir.

Anahtar kelimeler: *Kikuchi-Fujimoto, Histiositik Nekrotizan Lenfadenit, Lenfadenopati*

INTRODUCTION

KFD is a rare disease characterized by a benign course, which had been first defined in Japan almost simultaneously by Kikuchi and Fujimoto in 1972 [1,2]. It is frequently seen in young women in their twenties and associated with autoimmune diseases like systemic lupus erythematosus, local lymph node enlargement and fever [3]. Reported cases are mostly from far eastern countries and there are only a few cases encountered in the world [4] and our country [5]. Definitive diagnosis is elicited by histopathological examination of lymph nodes [6]. We want to draw attention to the differential diagnosis of many other diseases that cause lymphadenopathy, which may be confused with this disease; causing unnecessary and wrong treatment for the patient.

CASE PRESENTATION

A 34 years old Turkish female was admitted to our clinic with painful right supraclavicular lymphadenomegaly. She was caucasian, weighted 55 kg, 165 cm tall and was working as a secretary. Patient has used various nonsteroidal antiinflammatory drugs for the tenderness at her neck which began 20 days ago. Right supraclavicular mobile lymphadenopathy of approximately 2 cm diameter was palpated on physical examination. The pati-

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ent didn't have high fever and neutropenia (Neutrophile count: 2.58) but elevated sedimentation (93 mm/h) was present. ASO, CRP, AST, ALT levels and other complete blood count parameters were within normal limits. Posteroanterior chest radiograph and computed tomography (CT) of the chest was obtained and bilateral two millimetric size supraclavicular lymph nodes were reported (Figure 1). Patient was non-smoker, had a history of Hashimoto's thyroiditis 3 years ago and had a daughter having Mobitz type 2 AV block.

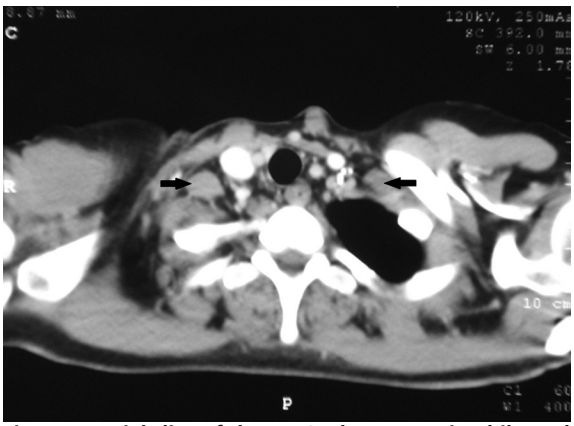

Figure 1. Axial slice of thorax CT demonstrating bilateral lymph nodes.

Right cervical lymph node excisional biopsy was applied to the patient under local anesthesia. Pathological result of the specimen was reported as KFD (Figure 2a and 2b) Patient received oral diclofenac potassium for two weeks and regularly controlled at outpatient clinic. Her symptoms disappeared in second week and no recurrence or extra symptoms were detected at the end of 15 months follow-up.

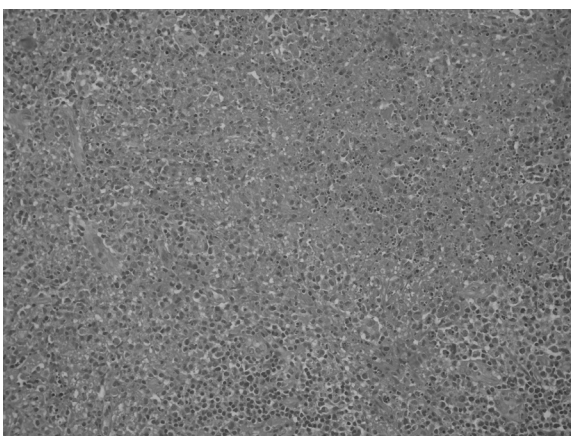

Figure 2a. Areas of karyorrhexis and karyolysis (H&Ex200).

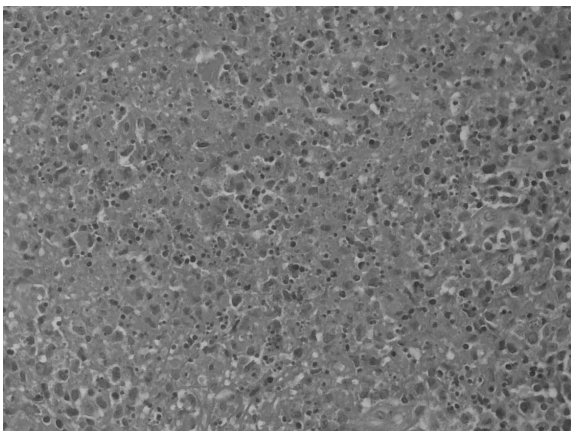

Figure 2b. Necrotic areas, karyorrhectic debris and scattered fibrin deposits (H&Ex40).

DISCUSSION

Although the pathogenesis of KFD is not fully understood, it is suggested that viral agents, hyper immunity or autoimmunity triggered by different antigens and cellular apoptosis are involved in etiology. Researchers mainly focused on viral etiology, particularly Epstein-Barr virus, Cytomegalovirus and Human herpesvirus 6 have been widely investigated [7]. KFD pathogenesis have common points with Kaposi's sarcoma-associated human herpesvirus 8 (KSHV/HHV 8) [8]. Also, enterocolitica serogrup 9 and 3, toxoplasma gondii, human parvovirus B19 infections are found in patients with KFD [9]. But, so far the specific pathogenic factor has not been definitively identified.

Some authors emphasize the role of immunological mechanisms involved in the pathogenesis of KFD and consider this disease as a rare manifestation of systemic lupus erythematosus (SLE). Previous literature on KFD frequently addressed the link between KFD and SLE, and the reported rate was 1.3-7% in the population of KFD patients [10,11]. Numerous studies have also shown an association between KFD and a wide spectrum of other autoimmune diseases, including Hashimoto's thyroiditis, polymyositis, mixed connective tissue disease, Still's disease, autoimmune hepatitis, and antiphospholipid syndrome [12-14].

The clinical and microscopic features of KFD

may closely resemble lymphoma as Kikuchi's disease shows preservation or only partial effacement of the lymph node architecture with patent sinuses, active histiocytes, and a low mitotic rate and it lacks a starry-sky pattern of macrophages [15]. Karyorrhectic debris was seen in our case. However, lymphoma has been ruled out with absence of atypical cells. Cat-scratch disease presents with necrosis including neutrophils and palisading histiocytes and contact with cats, but absence of these findings helped us to make differential diagnosis.

Laboratory findings include leukopenia, elevated transaminases and increased erythrocyte sedimentation rate. Sedimentation increase is usually mild or medium degree. Atypical lymphocytes may be seen on peripheral blood smear [16]. KFD is usually self-limiting, although rapidly advancing and even rare fatal cases had been reported. Recurrence was reported with ratio of 3-4 %. Some cases even relapsed with cervical swelling and fever after 28 months from first admission. Excisional or incisional biopsy of lymph node is required for definitive diagnosis. Sensitivity of lymph node aspiration cytology was reported as 56.25 % [17]. Usually the disease is completely cured with surgical resection as in our case [18].

CONCLUSION

Diagnosing KFD is important. Not just because of the wrong diagnosis rate of 40%, but also because of the severe diseases like non-Hodgkin lymphoma, SLE and Cat scratch disease which can lead to confusion in differential diagnosis. Kikuchi's disease should always be considered in the differential of cervical lymphadenopathy especially diagnosis in young females, to save the patient from unnecessary investigations and treatment.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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