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Kikuchi-Fujimoto disease in a young female with discoid lupus and alopecia

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Case Report

Abstract

Kikuchi-Fujimoto disease is an uncommon, benign, autoimmune condition characterized by lymphadenopathy, fever, and neutropenia. It is a self-contained condition of unidentified etiology. A 38-year-old female of Iranian Kurdish origin referred to us with multiple neck swellings, fever, and generalized arthropathy. There was no weight loss and no history of tuberculosis, medication intake, or allergy. Clinical examination revealed bilateral large, mobile, and tendercervical lymphadenopathy. Cardiovascular, respiratory, and neurological examination was normal. The patient had leucopenia and thrombocytopenia. Kikuchi-Fujimoto disease must be considered in the differential diagnosis of a female patient with fever and cervical lymphadenopathy.

KEYWORDS: Kikuchi-Fujimoto Disease, Lymphadenopathy, Alopecia

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Introduction

A benign and rare disease named Kikuchi-Fujimoto disease (KFD) was described for the first time by Kikuchi and Fujimoto independently in 1972.[1] Currently, the disease is well recognized. It is an uncommon cause of lymphadenopathy, mainly affecting young women. In spite of many reports and studies, the etiology of this disease is indistinct. Some initial reports hinted at Yersinia enterocolitica and Toxoplasma gondii as possible causative agents of KFD; however, viral or autoimmune factors may also be involved in the disease process.[2] The disease manifests itself with cervical lymphadenopathy, fever, or flu-like symptoms. Malaise, weight loss, loss of appetite, nausea, vomiting, diarrhea, chest pain, splenomegaly, and hepatomegaly are also included in the presentation of the disease. Laboratory tests include having elevated erythrocyte sedimentation rate (ESR), anemia, neutropenia, and lymphocytosis.[3]

The diagnosis of KFD is made through histopathologic features of the lymph node. On the other hand, morphological features may suggest systemic lupus erythematosus (SLE), non-Hodgkin lymphoma (NHL), and reactive lymphadenopathy. Therefore, for the differential diagnosis of KFD, it is appropriate to attempt to heighten awareness of the disease, especially in otolaryngologists and experienced pathologists.[4-6]

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Case Report

A 38-year-old female of Iranian Kurdish origin referred to us with multiple neck swellings, fever, and generalized arthropathy. There was no weight loss, and no history of tuberculosis or contact with patients with tuberculosis. The patient had no history of any medication intake, allergy, or any other background medical conditions, except for a history of renal stone and dysuria. Clinical examination revealed bilateral, large, mobile, and tender cervical lymphadenopathy; the larger nodes being the left supraclavicular lymph node which measured about 19 × 5 mm as well as axillary lymph nodes which measured about 24 × 7.5 mm. Lymph nodes were not palpable in other parts of the body. The patient's cardiovascular, respiratory, and neurological examination was normal. The abdomen was tender with normal bowel sounds. Throat examination was also normal. Routine hematological parameters, like hemoglobin, complete blood count, and peripheral smear, showed leucopenia and thrombocytopenia (white blood cell count = 2320 mm^3, lymphocyte = 30%, neutrophils = 53%, platelets = 113). High lactate dehydrogenase (LDH) (1202) with lymphadenopathy was suggestive of potential malignancy. ESR was 20 mm/1st hr. Blood glucose, urea, creatinine, and sodium levels were normal. Results showed normal morphology for lymphocytes with no lymphocytosis. Biopsy was taken from cervical lymph nodes. Pathology results revealed 3 clinical impressions, including lichen planopilaris, discoid lupus erythematosus, and alopecia areata; however, scalp lesion excision was compatible with discoid lupus erythematosus.

Liver function tests were normal. Blood and urine cultures were negative. Abdominal ultrasound and chest radiograph was normal. Staining for acid-fast bacilli (AFB) was also negative in the biopsy take from the lymph node as well as the sputum. In spite of 1 week antibiotics treatment, the patient continued to have fever and persistent lymphadenopathy; therefore, lymph node biopsy was obtained from the patient. Histological features suggested necrotizing lymphadenitis consistent with Kikuchi-Fujimoto lymphadenitis (Figure 1). Skin examination showed discoid lesions and the patient suffered from hearing loss. Additionally, diagnosis of the disease was confirmed by 3 pathologists and 3 oncologists.

The patient was treated symptomatically with nonsteroidal anti-inflammatory drugs (NSAIDs) resulting in regression of the lymph nodes. Follow-up of the patient showed improvement, and after 2 years of follow-up, our patient remained asymptomatic.

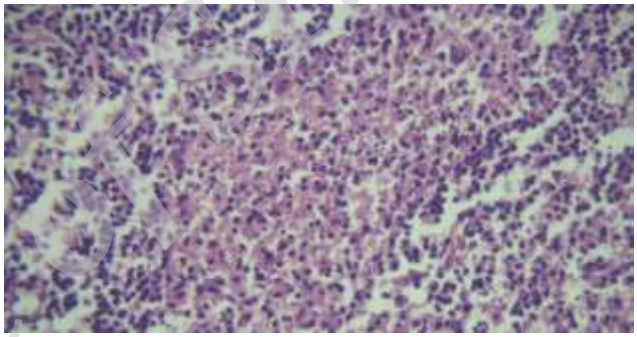

Figure 1. The affected nodes show focal well circmscribed paracortical necrotizing lesions. There are aboundant karyorrhectic debries, scattered fibrin deposits and collection of mononuclear cells. Plasma cells and neutrophils are very scanty a feature of diagnostic importance. The most important differential diagnosis is with malignant lymphoma with secondary necrosis.

Discussion

The presence of lymphadenopathy always raises concern about cancer. Patients with palpable lymph nodes in the supraclavicular region are at high risk for cancer.[3] Localized lymphadenopathy is common, and 55% of all cases involve lymph nodes in the head and neck, and the cervical region. In contrast, 1% of all cases of lymphadenopathy are attributed to supraclavicular lymph nodes, 5% to axillary nodes, and 14% to inguinal nodes.[3] KFD is often diagnosed by cervical tender lymphadenopathy accompanied by fever, and upper respiratory tract symptoms. Less common symptoms include

arthralgia, skin rash, weakness, and night sweats.[3] Other complications include weight loss, diarrhea, anorexia, chills, nausea, vomiting, chest and abdominal pain, and hepatosplenomegaly.[7] No definite cause for the disease has been recognized as yet; however, viral agents, such as Epstein-Barr virus (EBV), human immunodeficiency virus (HIV), herpes simplex virus, dengue virus, human T-lymphotrophic virus (HTLV), and parvovirus B19, have been suggested as possible etiological agents.[8] Toxoplasma and other bacterial agents, like Yersinia enterocolitica, Bartonella, and Brucella, have also been considered. KFD sometimes presents in conjunction with SLE; thus, the involvement of autoimmune mechanism has also been suggested.[2] There are several reports suggesting an association between KFD and SLE. However, no convincing evidence is available to confirm this relationship and the pathogenesis of KFD is unclear.[2]

Clinically, KFD may mimic SLE or lymphoma (especially T-cell non-Hodgkin lymphoma) as both these diseases can present with lymphadenopathy and fever, and the skin lesions of patients with KFD can resemble those seen in patients with SLE. Therefore, Histopathologic examination will help us distinguish KFD from other diseases.[2]

The histological feature which helps in the differentiation of KFD from lymphadenopathy of SLE is almost total absence of plasma cells in the involved nodal tissue. Moreover appropriate serologic tests should be conducted to exclude SLE. Features that distinguish KFD from malignant lymphoma include incomplete architectural effacement with patent sinuses, presence of numerous reactive histiocytes, relatively low mitotic rates, absence of Reed-Sternberg cells.[9]

No specific treatment is available for KFD as yet. Treatment is generally symptomatic. nonsteroidal anti-inflammatory drugs (NSAIDs) may be used to alleviate lymph node tenderness and fever.[10] Use of corticosteroids has been recommended in severe forms of the disease. Intravenous immunoglobulin has also been tried with relative success. The disease usually runs a benign course and the condition is self-contained, usually subsiding after several weeks to months with a recurrence rate of 3 to 4%.[9] The disease is an uncommon cause of lymphadenopathy. Because patients with lymphadenopathy refer to different physicians, the disease should be recalled and ruled out. The distinctive feature of our case was the diagnosis of discoid lupus together with alopecia.

Conclusion

KFD is rare; however, when a young female patient refers to us with fever and cervical lymphadenopathy KFD must be considered among the differential diagnosis. Clinically, the disease may resemble lymphoma or SLE. Therefore, a careful histopathological examination is essential for an accurate diagnosis. Moreover, early diagnosis of the disease is crucial to avoiding costly, painful, and pointless evaluations or treatments.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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