Kikuchi-Fujimoto disease in a 11 year male child : A Rare Case report and Review of literature

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ABSTRACT

Kikuchi-Fujimoto disease (KFD) is a rare, benign disease of unknown etiology and mainly involves cervical lymph nodes. Clinically it may mimic tuberculosis or lymphoma and cause difficulty in diagnosis. Accurate diagnosis of Kikuchi-Fujimoto disease (KFD) requires histopathological examination. Herein we report a case of an 11-year-old male child presented with right sided posterior cervical lymphadenopathy with low grade fever and who was clinically misdiagnosed as tuberculosis. Excisional biopsy was performed and histopathology confirmed the diagnosis of Kikuchi-Fujimoto disease (KFD). He was given symptomatic treatment and his recovery was uneventful. Kikuchi-Fujimoto disease (KFD) affects mostly young adult females, but rarely may affects children. We present this case due to its rare age group presentation.

Key words: Kikuchi-Fujimoto disease, cervical lymph nodes

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INTRODUCTION

Kikuchi-Fujimoto disease (KFD) is also known as Kikuchi disease, histiocytic necrotizing lymphadenitis, Kikuchi necrotizing lymphadenitis, phagocytic necrotizing lymphadenitis, sub acute necrotizing lymphadenitis, and necrotizing lymphadenitis.[1] Kikuchi-Fujimoto disease (KFD) is an uncommon and idiopathic cause of cervical lymphadenitis. It was first described in Japan by Dr Masahiro Kikuchi in 1972 and independently by Y. Fujimoto.[2,3] It is a rare, benign and self-limiting disease, mainly affects the young adults and predominantly more common in females as compared to males with male female ratio of 1:4.[4] So awareness of this disease in clinicians and pathologists might

help to prevent misdiagnosis and inappropriate treatment. We report a case of Kikuchi-Fujimoto disease that occurred in a 11-year-old male child who was clinically misdiagnosed as tuberculosis.

CASE REPORT

An 11 year-old male presented with multiple swelling in right side of neck since 4 weeks. He also complained of low grade intermittent fever with cough, anorexia and weight loss. He did not have any history of chest pain or breathlessness. No history of rash, arthralgia, joint pain, night sweats. There was no history of tuberculosis and similar swelling in past. But his father had taken treatment of tuberculosis in past. On examination he had multiple enlarged lymph nodes in posterior cervical region. Lymph nodes were approximately 2-3 cm in size, mobile, firm, non tender and without any discharging sinus. Swellings were not fixed to underlying structure. Systemic examinations were unremarkable.

Laboratory investigation revealed Hb-10.2 gm/dl, TLC-3200/cumm with mild lymphocytosis. ESR-26 mm in 1st hr. Chest radiograph and ultrasonography (USG) abdomen were normal. RA factor, anti-nuclear antibodies, Paul Bunnel test and Mantoux test were negative. The patient was clinically diagnosed as tuberculous lymphadenitis and sent for cytological examination. Fine needle aspiration cytology of cervical lymph nodes was done which revealed reactive lymphadenitis and patient was given oral antibiotics but patient continued to have fever and lymphadenitis. Finally excisional biopsy of the cervical lymph node was done and sent for histopathological examination. Gross examination showed multiple matted lymph nodes measuring 4x3.5x1cm. Externally capsulated and irregular surface. Cut section showed multiple matted lymph nodes largest 2cm in diameter, yellowish white in colour with few brownish areas [Figure 1].

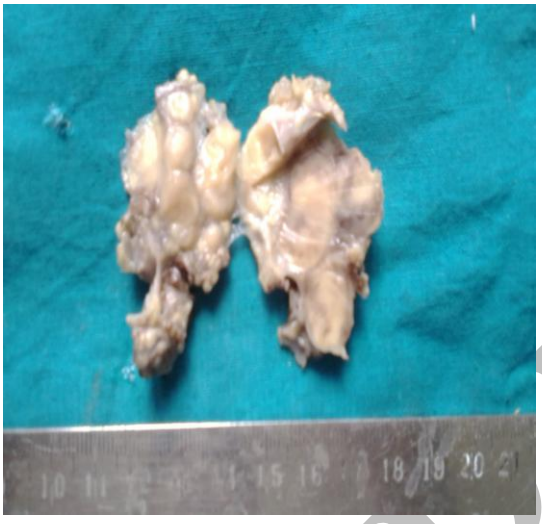

Figure 1: Cut section showed multiple matted lymph nodes largest 2cm in diameter, yellowish white in colour with few brownish area.

Histologically, serial sections showed focal well circumscribed paracortical coagulative necrosis with karyorrhectic debris. The necrotic foci were surrounded by reactive histiocytes, plasmacytoid monocytes, immunoblasts, small and large lymphocytes. Few areas showed lymphoid follicles with germinal centres. Few vessels showed fibrinoid necrosis. No evidence of granuloma or malignant cells [Figure 2 & 3].

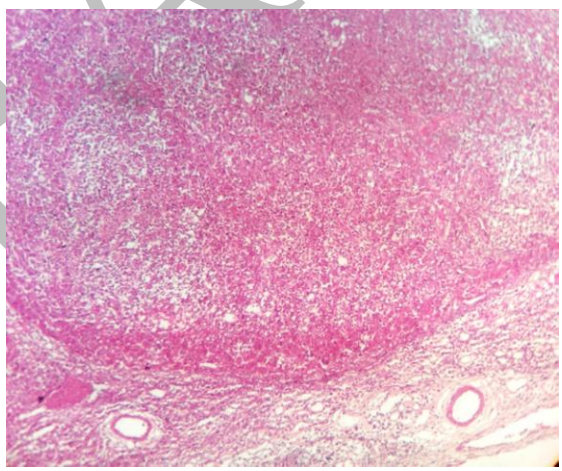

Figure 2: serial sections showed focal well circumscribed paracortical coagulative necrosis with karyorrhectic debris (H & E X100)

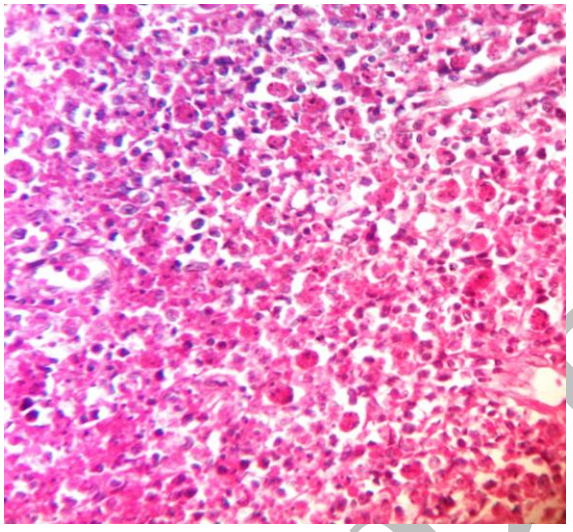

Figure 3: The necrotic foci were surrounded by reactive histiocytes, plasmacytoid monocytes, immunoblasts, small and large lymphocytes. (H & E X400)

Ziehl Neelsen stain revealed no acid fast bacilli. The histological findings confirmed the diagnosis of Kikuchi-Fujimoto disease (KFD). He was managed with symptomatic treatment and had complete resolution of lymph nodes.

DISCUSSION

Kikuchi-Fujimoto disease (KFD) is a rare and benign disease, mainly affects the young adults with male female ratio of 1:4.[4] So awareness of this disease in clinicians and pathologists might help to prevent misdiagnosis and inappropriate treatment.

Kikuchi-Fujimoto disease (KFD) has a worldwide distribution especially in Japanese.[5] Cervical lymph nodes are involved in 80%, axillary lymph nodes in 14% and 12% in supraclavicular lymph nodes.[6] Involvement of mediastinal, peritoneal and retroperitoneal regions is uncommon.[7] Clinically patient present with lymphadenopathy with low grade fever and may be associated with malaise, nausea, weight loss, gastrointestinal upset, myalgia, arthralgia. Few patients may have hepato-splenomegaly and skin rash.[8]Our patient presented with posterior cervical lymphadenopathy and above similar symptoms were observed except gastrointestinal upset, arthralgia, hepato-splenomegaly and skin rash. Atypical presentation of Kikuchi-Fujimoto disease causing meningitis has been reported by Noursadeghi M.[9]

The etiology of the disease is not clear. Mostly the microbial/viral agents such

as Epstein barr virus (EBV), Human immunodeficiency virus (HIV), Herpes simplex virus, dengue virus, Human T lymphotrophic virus 1 (HTLV1), Parvovirus B19, Toxoplasma and other bacterial agents like Yersinia enterocolitica, Bartonella, Brucella have been suggested. An autoimmune mechanism has also been proposed because 40 % of patients of Kikuchi-Fujimoto disease (KFD) may be associated with systemic lupus erythematosus (SLE) hence patients with these condition should be followed long term for early detection of SLE.[10] D. Belder-Preston reported a case of atypical KFD with retroperitoneal lymph node involvement and several features mimicking systemic lupus erythematosus. [11]

The pathogenesis of Kikuchi-Fujimoto disease (KFD) is still not fully understood. It is now proposed that Kikuchi-Fujimoto disease (KFD) is a non-specific hyper-immune reaction to a variety of infectious, chemical, physical and neoplastic agents and results in activation of T lymphocytes and histiocytes. Proliferating T cells undergo apoptosis, leading to multiple paracortical areas of coagulative necrosis within the lymph nodes and then removal of nuclear debris by histiocytes .[12]

No laboratory or radiological tests are available that confirmed the diagnosis of Kikuchi-Fujimoto disease (KFD). However complete blood count, X-ray chest, USG abdomen, USG/CT neck, antinuclear antibody, C-reactive protein may be done to exclude other conditions. FNAC also has a limited role in diagnosis of Kikuchi-Fujimoto disease (KFD) hence excisional biopsy is only confirmatory test.[13] The characteristic histopathological findings of Kikuchi-Fujimoto disease (KFD) are multiple paracortical areas of coagulative necrosis with abundant karyorrhectic debris, incomplete distortion of the nodal architecture, and large number of different types of histiocytes at the margin of the necrotic area. These karyorrhectic foci are formed by histiocytes, plasmacytoid monocytes, immunoblasts, small and large lymphocytes, mainly CD8+ T lymphocytes. Neutrophils are typically absent and plasma cells are absent or scarce.

The differential diagnoses of Kikuchi-Fujimoto disease (KFD) are tuberculosis, lymphoma, SLE and viral/bacterial lymphadenitis, infectious mononucleosis and cat-scratch disease. Absence of epitheloid granuloma, negative Mantoux test and negative Ziehl Neelsen

stain ruled out tuberculosis in our case. Negative anti-nuclear antibody (ANA) and anti-dsDNA antibody as well as characterstics histomorphological features suggestive of KFD ruled out SLE in our case. The absence of granulocytes in areas of necrosis and lack of follicular hyperplasia differentiate these cases from lymphadenitis caused by cat-scratch disease and other bacterial infections. Early stage lesions without overt necrosis may be distinguished from lymphoma by a) focal nature of the lymph node involvement with incomplete architectural effacement b) the presence of plasmacytoid monocyte at the periphery of the lesion and c) bland cytologic features of the mixed cells population in areas of involvement .[14]

Based on histopathologically, Kuo described three types of Kikuchi's disease: Proliferative, necrotizing, and xanthomatous. The proliferative features are seen in about one-third of cases and have an inflammatory infiltrate. Half of the cases show necrotizing pattern and the xanthomatous type is rare and has abundant foam cells.[15] Kikuchi-Fujimoto disease (KFD) is a self-limiting disease and resolves spontaneously over weeks to months. There is no specific treatment of Kikuchi-Fujimoto disease (KFD). Only symptomatic treatment like analgesic, antipyretics and NSAID may be given and in severe cases corticosteroid may be used. Recurrence rate of the disease has 3-4% reported.[16] Our patient was managed with symptomatic treatment and recurrence did not occur.

CONCLUSION

Kikuchi-Fujimoto disease (KFD) is an uncommon, self limiting disease and a rare cause of cervical lymphadenopathy. Careful clinical history, laboratory investigation and histopathological examination are mandatory for every case of cervical lymphadenopathy which will help in accurate diagnosis of disease and to minimize unnecessary evaluation and treatment.

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