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Case Reports

Kikuchi-Fujimoto Disease: A Case Report and Review of literatures

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Introduction

Kikuchi-Fujimoto Disease (K-FD), or histiocytic necrotizing lymphadenitis, was first described in 1972 simultaneously by Kikuchi et al[1] and Fujimoto et al.[2] It is a benign and self-limited disorder, characterized by regional cervical lymphadenopathy with tenderness, usually accompanied by mild fever and night sweats.[3] Kikuchi-Fujimoto disease is an extremely rare disease known to have a worldwide distribution with higher prevalence among Japanese and other Asiatic individuals.[3] Recently a young girl was admitted with fever and lymphadenopathy and being diagnosed as Kikuchi- Fujimoto disease and documented here for future references.

Case Report

A 10 year old girl, was admitted on17.09.2014 in the Department of Paediatrics of Dhaka Medical College Hospital with fever for 20 days. It was irregular in nature, low grade during daytime and high grade at night, associated with chills and rigor, subsided with antipyretic. Fever was also accompanied by non-productive cough and loss of appetite. Along with fever and cough, parents also noticed a painful swelling on the right-side of neck from the first day of fever. The girl was evaluated and treated locally but her condition didn't improve. Finally she was admitted in Dhaka Medical College Hospital for further evaluation and m1anagement. She had no history of contact with TB patient, scratching, rash, photo-sensitivity, joint or abdominal pain, haematuria or, other form of bleeding manifestations. She was immunized as per EPI schedule. Her past and family history were unremarkable. Clinical examination revealed that she was ill looking and co-operative, febrile (temperature-10ºF), mildly pale, respiratory rate 28/min, blood pressure 100/70mmHg.The right posterior cervical group of lymph nodes were enlarged, measuring about 0.5×0.3cm to 2.0×1.0 cm, tender, firm in consistency, not fixed with underlying and overlying structures, overlying skin was normal & there was no discharging sinus. BCG mark was present. She was non icteric, non-oedematous, bony tenderness absent, no rash or bleeding manifestation was observed over the skin. Other systemic examinations revealed normal. CBC-revealed leukopenia (3,500/Cumm),Hb-10.3gm/dl, plateletcount-2,50,000/ cumm,ESR-60 mm in 1st hr. Urine analysis was normal. Blood and urine culture revealed no growth, Febrile antigen-negative. Immunological and rheumatological tests e.g. ANA, anti-ds DNA, RA factor, Serum C3 and C4 were also normal. MT was negative and chest radiograph was also normal. Excisional biopsy of enlarged right cervical lymph node was done and histopathology revealed many apoptotic bodies and many histiocytes. No granuloma or malignant cells was seen. The findings was consistent with acute histiocytic necrotizing lymphadenitis (Kikuchi disease).She was initially treated by paracetamol bed rest and others supportive measures. The girl was discharged when her fever was subsided and advised for follow up.

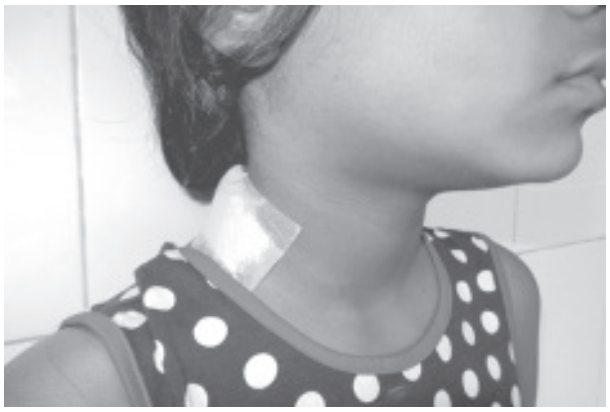

Fig.-1: *Photograph of the patient with dressing over the lymphnode excision area.*

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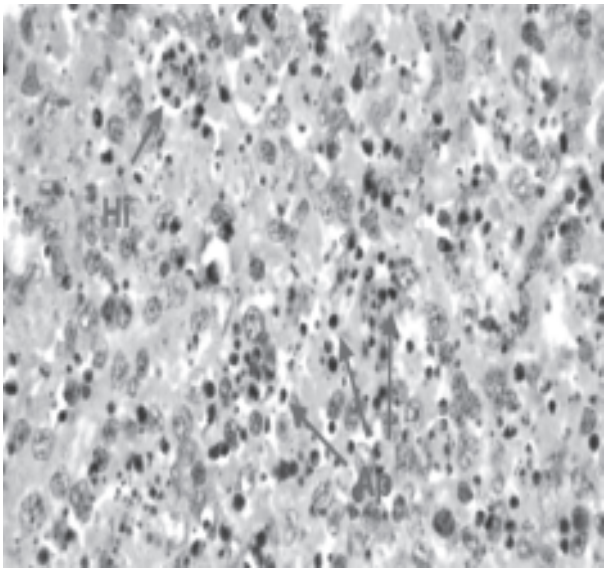

Fig.-2: *Section of lymph node revealed accumulation of many apoptotic bodies and histiocytes without presence of granuloma or malignant cell, characteristics features of Kikuchi-Fujimoto disease.*

Discussion

Kikuchi-Fujimoto Disease(K-FD) was originally reported in young Japanese female by Kikuchi et al[1] and Fujimoto et al[2] 1972. The condition has been described in both genders and in a variety of ethnic groups.[4] Women are affected more often than male.[5] The onset of K-FD is usually acute or sub-acute. It typically manifests as cervical lymphadenopathy with low grade fever in a previously healthy young individual. The enlarged lymph nodes, ranging from 0.5 to 4 cm in size, are tender and painful. Cervical lymph nodes are affected in around 80% of cases. Posterior cervical lymph nodes are affected in 65% to 70% of cases[6,7], other affected sites are axillary(14%) and supraclavicular (12%) nodal chains.8 Generalized lymphadenopathy occurs in a minority(5%) of cases. In our patient only right posterior cervical chain was involved. Extra nodal involvement in K-FD is uncommon, but cutaneous lesions has been documented in the literatures.[9,10] The skin lesions are most frequently located on the face or upper body and includes erythematous papules, plaques, indurated lesions, and ulcers. None of the lesions are pathognomonic. In rare cases, hepatomegaly and splenomegaly have been described in literatures. [11] Fever usually of low grade and present among 33-50% of patients.[12] It is usually associated with upper respiratory symptoms and typically lasts for 1 week, although it could persist for up to 1 month in rare cases.[12] Our patient has fever which was low grade at daytime and high grade at night. Other uncommon symptoms includes fatigue, joint pain, nausea, vomiting and sore throat. Night sweats and weight loss, the systemic symptoms frequently seen in lymphoid malignancy, are rare but have been reported.[5,11] Our patient had anorexia and weight loss. Leukopenia can be found in up to 50% of cases. Atypical lymphocytes in the peripheral blood have also been reported. Although the disease has been linked to SLE, as well as to other autoimmune conditions,[13,14] the real strength of such associations remains to be clarified.

K-FD is generally diagnosed on the basis of an excisional biopsy of affected lymph nodes. The diagnosis of K-FD merits active consideration in any nodal biopsy showing fragmentation, necrosis and karyorrhexis, especially in young individuals presenting with posterior cervical lymphadenopathy.[3] The main diagnostic problem encountered by the histopathologist is to distinguish Kikuchi disease from non-Hodgkin's Lymphoma.

Treatment is symptomatic (analgesics-antipyretics, non-steroidal anti-inflammatory drugs and rarely corticosteroids).Spontaneous recovery occurs in 1 to 4 months. Patients with Kikuchi-Fujimoto disease should be followed-up for several years to survey the possibility of the development of systemic lupus erythrometosus.[3] Our patient was treated with paracetamol, bed rest and followed up without any problem.

Conclusion

Although few cases were reported in Bangladesh but the prevalence of Kikuchi-Fujimoto disease is still unknown in Bangladesh. Cases may be present but remain unreported in our country. The disease usually benign in nature but sometimes it may appears as a malignant disease and require long term follow-up. Early recognition will reduces unnecessary evaluations, treatment as well as parent's anxiety. So when a patient comes with painfull cervical lymphadenopathy along with fever, clinician may consider Kikuchi Fujimoto disease as one of the differential diagnosis.

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