KIKUCHI FUJIMOTO DISEASE, A GREAT IMITATOR: A CASE REPORT OF A 14 YEAR OLD FEMALE PRESENTING WITH FEVER AND LYMPHADENOPATHY

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ABSTRACT

Kikuchi Fujimoto disease (KFD) is a rare disease of unknown aetiology, with higher prevalence amongst Japanese and east Asian population. It is a benign disease, runs in a limited course, characterized by necrotizing histiocytic lymphadenitis mostly affecting young women. KFD clinically presents with enlarged lymph nodes plus fever. Cervical lymph nodes are commonly affected but rare cases of generalized lymphadenopathy and involvement of retroperitoneal lymph nodes have been noticed too. Its recognition is significant as it can be mistaken for immunological, infective and even lymph-proliferative disorders like lymphomas thus preventing the patients from being subjected to costly diagnostic work up and treatment. Once a true diagnosis has been made the treatment of the disease mainly comprises of supportive measures. This is different from the management of other conditions which may be confused with it i.e. lymphoma, tuberculosis.

Keywords: Histiocytic necrotizing lymphadenitis, Kikuchifujimoto disease, Lymphoproliferative disorders.

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INTRODUCTION

Kikuchi Fujimoto disease (KFD), otherwise called histiocytic necrotizing lymphadenitis is an uncommon, idiopathic disease affecting Japanese and east Asian individuals more commonly with a male to female ratio of 1:4.[1-5].

The pathogenesis is still poorly understood but it is thought to be due to hyper immune reaction induced by various antigens or auto immune reaction in which apoptosis plays an important role. The most common signs and symptoms include fever, lymphadenopathy, fever, malaise, anorexia and raised inflammatory markers[6]. A definite diagnosis is made on the basis of histopathological assessment of the involved lymph nodes which reveals characteristic findings[7,8]. The disease is of benign nature having a self-limiting course with an outstanding prognosis and a magnificent response to therapy. The management of the disease is mainly based on the supportive measures with analgesics and antipyretics. However steroids and other immunosuppressive agents have shown promising roles in few complicated cases[9,10].

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CASE REPORT

A 14 year old girl, known case of dilated cardiomyopathy (DCMP) since 3 years, taking medica-tions for DCMP, presented to us with fever, malaise and enlarged cervical lymph nodes. Her family history was negative for auto-immune diseases. She took paracetamol for 3 weeks with no clinical response. Physical examination revealed fever of 39˚C, she had tender, mobile bilateral cervical lymphadenopathy. Moreover she also had positive fluid thrill and mild pedal edema.

The laboratory evaluation revealed a haemoglobin (Hb) of 10.8 gm/dl, MCV 69 fl; platelets, 189000/cmm; and leucocytes 13200/cmm. The ESR was 49mm/1st hour. The peripheral smear was consistent with anemia of chronic disease with normocytic picture. The biochemical investigations revealed normal liver biochemistry and renal functions. Test for anti-nuclear antibody was negative. Serum LDH was 935 IU/L. The cervical ultrasound revealed enlarged hypo dense lymph nodes bilaterally. The chest-abdomen CT demonstrated minimal bilateral pleural effusion, moderate ascites, there was further an evidence of enlarged cervical and

mediastinal lymph nodes with lymphadenopathy seen in para aortic and retroperitoneal regions too. The D/Ds of lymphoma vs tuberculosis (TB) were suggested by the radiologist and the same two differentials were made provisionally too on the basis of under lying symptoms, lymphadenopathy, polyserositis and raised LDH levels. An excisional cervical lymph node biopsy for histopathological evaluation was performed which revealed acute and chronic inflammation with areas of necrosis and karyorrhexis debri with no evidence of granuloma formation of malignant cells. Final diagnosis of KFD was established on the basis of characteristic histopathological findings.

The patient was symptomatically treated with NSAIDS and anti pyretics. She was also given oral prednisolone therapy 30mg per day. Her fever started to settle 24 hours after starting steroids therapy and she showed clinical improvement too. The size of the lymph nodes also regressed at one month follow up and her symptoms resolved as well. The patient was switched to tapering doses of prednisolone.

DISCUSSION

KFD is a benign disorder with a self-limited course that affects females more commonly under the age of 30 years[6].

The clinical manifestations of fever, malaise, lymphadenitis in a young woman is certainly suggestive of an infectious or an autoimmune disease. KFD has been reported to be an early stage of SLE. The infectious etiology comprises of a hyper immune reaction in a genetically predisposed individuals whereas autoimmune etiology could be attributable to significant correlation between KFD and autoimmune diseases[2].

No specific diagnostic tests are available; Although some patients have an elevated ESR. Definite diagnosis is established by histopathological findings of lymph node biopsy. The characteristic histopathological findings include irregular para cortical areas of coagulative necrosis and karyorrhectic debri with large number of histiocytes at the margin of necrotic areas[10].

KFD is a rare disease which most of times presents as diagnostic dilemma. It should be a part of differential diagnosis of lymph node enlargement as its therapy is significantly different from other causes, for example unnecessary chemotherapy and radiotherapy. Treatment is mainly supportive with NSAIDS and paracetamol mainly used to alleviate the tender lymphadenitis and fever[10]. Corticosteroids have promising role where supportive measures fail to abate the symtoms. Other immunosuppressive agents like azathioprine, Cyclosporin, hydroxychloroquine and immunoglobulins have proven successful in individual cases[1]. Follow up for an extended period of time is necessary as recurrence has been reported and it is also believed that KFD may be a precursor of SLE[10].

CONCLUSION

KFD is a self-limited disorder of benign nature. It is a great imitator of various sinister conditions. It can be easily mistaken for lymphoma, tuberculosis, ASOD and so on. Timely diagnosis of the disease is necessary to avoid unneeded and harmful evaluations and treatment.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

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