Kikuchi-Fujimoto Disease: A rare cause of PUO - Case Report

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ABSTRACT

Kikuchi’s disease or Kikuchi-Fujimoto Disease (KFD) is a rare, benign self-limited condition defined by fever with tender regional (most often cervical) lymphadenopathy. There are cases reported worldwide, scarcely among Europeans but more frequently among the Japanese and Asian population. Here we present a case of Kikuchi disease in a young South Indian male patient.

KEYWORDS: Histiocytic, Kikuchi, Kikuchi Fujimoto, Lymphadenitis

INTRODUCTION

A rare but important disease, namely Kikuchi Fujimoto disease is being reported here. The importance of this condition lies in the fact that the clinical presentation is most often fever and tender, enlarged lymph nodes. The diagnosis is based on the histopathology of the lymph node, and is characterized by histiocytic necrotizing lymphadenitis. While this disease is in fact most often benign, the differential diagnosis is constituted by far more serious ailments including lymphoma[1].

The treatment modalities of the two are obviously poles apart. In reporting this case, we attempt to highlight the importance of considering Kikuchi’s disease as an important differential diagnosis when dealing with a case of pyrexia of unknown origin, especially in India as the disease is prevalent in Asia. A notable feature in our case was the lack of clinically palpable lymph nodes which were picked up incidentally on ultrasonography of the neck. We have also done a concise review of literature to help validate our case.

CASE REPORT

A 25 year old male patient presented to the OPD with chief complaints of intermittent fever and dry cough since three days. Patient was prescribed Co-amoxiclav and antihistaminics and symptoms resolved. The patient returned to OPD one and a half months later with complaints of fever with non-productive cough for a month associated with history of a 4kg weight loss. On further questioning, patient confirmed no prior contact with a known or suspected case of tuberculosis, no history of drug intake, and no myalgia or rash. Clinical examination, both general and systemic were unremarkable except for a Temperature of 100°F. Patient’s initial investigations including Complete Blood Count, Renal Function Tests, Liver Function Tests, Urine Routine, Erythrocyte sedimentation rate (5mm/hr) and procalcitonin levels (0.25ng/ml) were all found to be within normal range. Sputum culture, Sputum for AFB, Peripheral smear for Malarial parasite, Blood Culture, Urine culture, Widal, HIV 1 and 2 serology were also ordered and found to be negative.

Patient was started on broad spectrum antibiotics (Cephalosporin) and antipyretics. Antibiotics were escalated on day 3 to Piperacillin-Tazobactam but fever spikes still persisted. Persistent fever spikes warranted further investigations, amongst which Paul-Bunnel, Weil-Felix tests and ANA profile were negative. 2D ECHO revealed no abnormalities. However, Mantoux test was strongly positive (17x20mm induration) and USG neck revealed enlarged cervical lymph nodes that were not clinically palpable. A lymph node excision biopsy was carried out and histopathological examination of the tissue revealed histiocytic infiltration with expanded paracortex and circumscribed areas of necrosis (Fig. 1, 2). Based on clinic-pathological correlation, a diagnosis of Kikuchi disease was made. All antibiotics
were stopped and patient was started on Prednisolone 20 mg OD. Fever spikes abated within 24 hours of starting steroids. Patient is currently being followed up regularly on an OPD basis and remains asymptomatic.

**DISCUSSION**

Kikuchi-Fujimoto Disease (KFD) or Kikuchi-Fujimoto lymphadenitis or Kikuchi’s disease is a rare lymphohistiocytic disorder first described in 1972 in Japan, by Dr Masahiro Kikuchi and Y. Fujimoto independently. It is one such condition which often produces confusion because of features which mimic life threatening conditions including lymphoma, tuberculosis and systemic lupus erythematosus[1]. Therefore, it remains an underdiagnosed condition.

KFD historically has a well documented Asian female preponderance (1:3) [2], though recent literature supports an almost equal male to female ratio(1:1.25) [3]. The etiopathogenesis of KFD is not well defined. Though multiple viruses and bacteria are suspected there is no consistent evidence. The presence of HLA class II genes are more frequent in patients with KFD. In particular, the incidence of DPA1*01 and DPB1*0202 [4] alleles is significantly higher in patients with KFD than in healthy control subjects. These genes are extremely rare or absent among Caucasians but relatively common among Asiatic people (e.g. French, 0.4%; Italian, 0.8%; Korean, 9.9%; and Japanese, 4.5%). This might provide an explanation for the epidemiology of the disease[5].

KFD present with tender, palpable lymph nodes, generally unilateral, mainly in the cervical region uncommonly in other nodal groups. Fever is low grade, seen in 30-50% of patients and associated with upper respiratory symptoms. Less frequent symptoms include weight loss, nausea, vomiting, sore throat, and night sweats. Extranaodal sites involved are mainly the skin (usually the face), bone marrow and deranged liver functions[6]. Skin involvement pattern in KFD is non-specific. It has been suggested that interface dermatitis in the presence of KFD could be a valuable marker of evolution of KFD to SLE[7] Laboratory investigations are essentially normal except for mild granulocytopenia. FNAC per se has a very limited diagnostic accuracy (56.3%) and excisional Lymph node biopsy is indicated in suspected cases[8].

In our case, features of PUO were prominent with weight loss but characteristically lacked palpable lymphadenopathy. Enlarged lymph nodes were incidentally picked up by ultrasonography of the neck. His Mantoux test was falsely positive[9] and nodal biopsy showed histiocytic infiltrate and abundant karyorrhectic debris which are hallmarks of KFD. On starting steroids he showed a rapid clinical improvement and was discharged. He is being regularly followed up in the OPD and continues to be asymptomatic.

**CONCLUSION**

KFD(Kikuchi Fujimoto disease), a clinicopathologic diagnoses, though a rare entity is easily amenable to treatment with minimal recurrences. Along with malignancies, autoimmune disorders and granulomatous diseases, it should form a part of differential diagnoses for PUO in endemic (Asian) population.

**Conflicts of interest:** NIL

**REFERENCES**

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