Case Report

Kikuchi-Fujimoto Disease, a rare cause of cervical lymph adenopathy.

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Abstract

Kikuchi-Fujimoto Disease (KFD) present with Dysphagia, fever and lymphadenopathy. A young adult Bangladeshi female presented with fever and cervical lymphadenopathy. Broad spectrum antibiotic (3rd generation oral cephalosporin) were given but no improvement after one month. FNAC suggested non specific lymphadenitis. Later on excision biopsy of cervical lymph node was done and histopathology suggested KFD. Oral prednisolone was given showing improvement and no relapse was en-counted. Awareness of this disorder by clinicians and pathologists will help misdiagnosis and inappropriate treatment.

Introduction

Kikuchi Fujimoto disease (KFD) is an enigmatic, benign & self limited syndrome characterized by regional Lymphadenopathy with tenderness, predominantly in cervical region. Usually accompanied by mild fever & night sweats. Initially described in Japan, KFD was first reported in 1972 almost simultaneously by Kikuchi1 and Fujimoto et al2 as lymphadenitis with focal proliferation of reticular cells accompanied by numerous histocytes and extensive nuclear debris3.

We present a case of KFD having cervical lymphadenopathy along with fever & weight loss. The review of literature showed only one case series of 58 KFD patients in southern Taiwan. Of these one patient had odynophagia4. No case of retropharyngeal LN enlargement has been reported5.

Case Report

Mrs. Shahida Begum, Age-29 yrs, Comilla Sadar, Comilla presented on March, 2013 with fever, swelling of the neck for 1½ months. She had no H/o tuberculosis exposure. Physical examination revealed enlargement of cervical lymph nodes. The LNS were multiple, 2–3 cm in diameter, firm in consistency, discrete, mobile, in Rt posterior triangle, Jugolodigastic & submandibular regions. Her temperature was 100°F, weight 59 kg, CBC shows ESR 45 mm no neutropenia, MT negative & normal CXR. Broad spectrum antibiotic 3rd generation oral cephalosporin was given but there was no improvement after 2 weeks. FNAC shows nonspecific lymphadenitis. Excision biopsy of cervical lymph node from posterior triangle was done. Histopathology revealed areas of necrosis & nuclear dust with surrounding infiltration of foamy histocytes. ANA and anti DS DNA were negative. A diagnosis of KFD was made. Oral prednisolone was given for 2 months in tapering dose started with 60 mg. daily. Her fever & swelling of lymph node started to disappear after 2 weeks. After two months neck swelling disappeared. The patient was seen after 6 months and there was no evidence of relapse of LN swelling & fever.

Discussion

KFD is an enigmatic, benign and self-limited syndrome characterized by regional lymphadenopathy with tenderness, predominantly in the cervical region, usually accompanied by mild fever and night sweats. Initially described in Japan, KFD was first reported in 1972 almost simultaneously by Kikuchi1 and Fujimoto et al2, as lymphadenitis with focal proliferation of reticular cells accompanied by numerous histocytes and extensive nuclear devris3. Kikuchi-Fujimoto disease is an extremely rare dis-

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ease known to have a worldwide distribution with a higher prevalence among Japanese and other Asiatic individuals\(^6\). Affected patients are most often young adults under the age of 30 years. The disease is seldom reported in children. Recent reports seem to indicate that the female preponderance was overemphasized in the past and that the actual ratio is closer to 1:1\(^7\).

There is much speculation about the cause of KFD; infection or autoimmune has been suggested. Some initial reports hinted at Yersinia enterocolitica and Toxoplasma gondii as possible causative agents of KFD, mainly on the basis of positive serologic test results. The role of Epstein-Barr virus (EBV), as well as other viruses, in the pathogenesis of KFD remains controversial. Nevertheless, the association between KFD and SLE has been reported with a frequency probably greater than that expected by chance alone\(^6\).

The onset of KFD is acute or sub acute, evolving during a period of 2 to 3 weeks. Cervical lymphadenopathy is present in 50% to 98% of cases, more commonly consisting of tender lymph nodes involving the posterior cervical triangle (88.5%), generally unilateral (88.5%). Lymph node size ranges from 0.5 to 4 cm (93.4%) and occasionally, lymph nodes are larger than 6 cm. Painful lymphadenopathy is seen in up to 59% of patients. Generalized lymphadenopathy has been reported in 1% to 22% of cases. Involvement of mediastinal, peritoneal and retroperitoneal regions is uncommon. In addition to lymphadenopathy, 30% to 50% of patients with KFD might have fever, usually low-grade, associated with upper respiratory symptoms. Less common manifestations include fever, axillary and mesenteric lymphadenopathy, splenomegaly, parotid gland enlargement, cutaneous rash, arthralgias, include fever, axillary and mesenteric lymphadenopathy, splenomegaly, parotid gland enlargement, cutaneous rash, arthralgias, myalgias, aseptic meningitis, bone marrow hemophagocytosis\(^9\). Our case presented with enlargement of Rt sided cervical lymph nodes? fever. Involvement of extranodal sites in KFD is uncommon but skin, eye and bone marrow being affected and liver dysfunction have been reported\(^6\). Kikuchi-Fujimoto disease is generally diagnosed on the basis of an excisional biopsy of affected lymph nodes. No specific diagnostic laboratory tests are available. Some patients have anaemia and a slight elevation of the erythrocyte sedimentation rate. Mild leukopenia has been observed in 25% to 58% of patients, whereas leukocytosis is found in 2% to 5% of cases. Moreover, 25% to 31% of patients have atypical peripheral blood lymphocytes\(^3,7\). The usefulness of fine-needle aspiration cytology (FNAC) to establish a cytologic diagnosis of KFD has been limited and in general it is less useful than excisional LN biopsy, the overall diagnostic accuracy of FNAC for KFD has been estimated at 56.3%\(^10\). Therefore excisional lymph node biopsy should be mandatory if clear-cut clinical and cytologic KFD finding are absent. Characteristic histopathologic findings of KFD include irregular paracortical areas of coagulative necrosis with abundant karyorrhectic debris, which can distort the nodal architecture and large number of different types of histiocytes at the margin of the necrotic areas.

Kikuchi-Fujimoto disease is typically self-limited within 1 to 4 months and possible recurrence rate of 3 to 4% has been reported\(^3\). Analgesics-antipyretics and nonsteroidal anti-inflammatory drugs may be used to alleviate lymph node tenderness and fever. The use of corticosteroids has been recommended in severe extranodal or generalized KFD but is of uncertain efficacy. In our case prednisolone was given to relives progressive lymphadenopathy, which showed rapid relief of symptoms.

**Conclusion:**
KFD is an extremely rare possibility to be kept in mind specially when dealing with a young female patient with fever and cervical lymphadenopathy. Awareness of this disorder, not only by clinicians, but pathologists might help prevent misdiagnosis and inappropriate treatment.
References:


