



International Journal of Medical Research & Health Sciences

www.ijmrhs.com Volume 2 Issue 3 July - Sep Coden: IJMRHS Copyright ©2013 ISSN: 2319-5886

Received: 6th Apr 2013 Revised: 2nd May 2013 Accepted: 8th May 2013

Case report

CERVICAL LYMPHADENOPATHY IN A YOUNG FEMALE: A CASE REPORT

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ABSTRACT

Kikuchi-Fujimoto disease is a rare disease of unknown etiology and characterized by benign self-limiting cervical lymphadenopathy. It is an extremely rare disease with higher prevalence amongst Japanese and other Asiatic individuals. Its recognition is important because it can be mistaken for immunologic, infective, and even lymphomas, and the patient may be subjected to inappropriate and costly diagnostic workup and treatment. A 21-year-old female presented to the clinic with chief complaint of swelling over the left neck with fever since 10 days. On examination, patient was febrile, and she had multiple small enlarged cervical lymph nodes, that were mildly tender. Other system examination was normal. Her laboratory investigations were normal. FNAC of the cervical lymph node was suggestive of necrotizing lymphadenitis. Patient was started on a course of ciprofloxacin. Her lymph node biopsy confirmed Kikuchi's disease. Immunohistochemistry further confirmed the diagnosis. The patient has been free of symptoms and in remission on followup after six months.

Keywords: Kikuchi-Fujimoto disease, necrotizing lymphadenitis, immunohistochemistry

INTRODUCTION

Kikuchi-Fujimoto disease, otherwise called histiocytic necrotizing lymphadenitis, is an idiopathic, rare disease generally presenting as cervical lymphadenitis. It was first described in Japan in the year 1972. It is reported to be more common in the Japanese and Asiatic individuals. The male to female ratio is reported to be around 1:4

CASE REPORT

A 21-year-old unmarried female presented to the clinic with a history of fever and multiple neck

swellings and with generalized myalgias and arthralgias of 10 days' duration. There was no other significant past medical history. The patient denied any history of previous Koch's or contact with any patient with Koch's. Her physical examination was significant in that she was febrile with a temperature of 100.8 degrees Fahrenheit. Her neck exam showed multiple, small lymph nodes, which were mobile and tender on the left side of the neck, the largest of which measured and 1.5 x 1.5 cm. There were no other lymph nodes palpated elsewhere in the

body. Her throat exam was normal with no pharyngitis or tonsillitis or tonsillar hypertrophy. Her cardiovascular, respiratory, and per abdomen exams were likewise normal. Patient's routine blood investigations including ESR were normal. She had normal electrolytes, liver function tests, and renal function tests. Mantoux did not show any induration. Her chest x-ray and abdominal ultrasounds were reportedly normal. She had ANA and anti dsDNA antibodies checked which were negative. USG of neck showed cervical lymphadenopathy involving the left upper and mid cervical nodes, the largest of which was 1.8 x 2.1 cms.

Patient was then subjected for FNAC of the cervical lymph node which was reported as showing necrotizing lymphadenitis, to rule out Kikuchi's disease. A lymph node biopsy was done which confirmed the diagnosis of Kikuchi-Fujimoto's disease, which was further confirmed with immunohistochemistry. The patient was treated symptomatically with ciprofloxacin and nonsteroidal anti-inflammatory drugs with resolution of her symptoms, and she has remained in remission after two years of follow up.



Fig.1: Smear from cervical lymph node showing reactive lymphoid cells with scattered crescentic histiocytes. Background shows necrosis with numerous tingible body macrophages.

DISCUSSION

Kikuchi-Fujimoto's disease remains an enigmatic condition of unclear etiology and one

that is rare except in the Japanese and Asiatic individuals and often leads to unnecessary investigations and potentially harmful treatments. The disease often presents with tender cervical lymphadenopathy in a female aged between 20 to 30 years. Though the etiology is still to be elucidated, viral infections with Herpes simplex virus, Epstein Barr virus, Parvovirus B19, and HTLV1 have been suggested, but this is controversial and not convincingly demonstrated¹. Other infectious agents which have been implicated in Kikuchi's disease include Brucella, Yersinia, Bartonella, and Toxoplasma. There are reports suggesting an association between Kikuchi's disease and SLE. It has been suggested by some authors that Kikuchi's disease may represent a self-limited autoimmune process which is induced by virus-infected transformed lymphocytes in a genetically susceptible individual².

Cervical lymphadenopathy is almost always the most common symptoms which bring the patient to physicians. The other less common symptoms include anorexia, nausea, vomiting, arthralgias, night sweats, and skin rash. It has also been reported as a cause of PUO³. Leukopenia and atypical lymphocytes are demonstrated in about 25% to 30% of the cases.

No specific laboratory tests are available; although some patients may have an elevated ESR Fine needle aspiration cytology has an overall diagnostic accuracy of around 56%⁴. Diagnosis is established by histopathologic findings of lymph node biopsy. The characteristic histopathologic findings include irregular paracortical areas of coagulative necrosis with abundant karyorrhectic debris, which distorts the nodal architecture, and large number of different types of histiocytes at the margin of the necrotic areas. The karyorrhectic foci are formed by different cellular types, predominantly histiocytes and plasmacytoid monocytes but also immunoblasts and small and large lymphocytes. Neutrophils are characteristically absent and plasma cells are

either absent or scarce. Of note, atypia seen in the reactive immunoblastic component is not uncommon and can be mistaken for lymphoma. The differentiation of KFD from SLE can sometimes pose problems because both can show similar clinical and histological features. Furthermore, KFD has been reported in association with SLE. Antinuclear antibodies (ANA) and anti-DNA antibodies were done in our patient and were negative. The immunophenotype of Kikuchi's disease is primarily composed of mature CD8-positive and CD4-positive T lymphocytes with very few B-lymphocytes. High rates of apoptosis are seen among lymphocytes and histiocytes. The histiocytes express histiocyte-associated antigens such as lysozyme, myeloperoxidase (MPO) and CD68 which can be detected by immunohistochemistry. Plasmacytoid monocytes are also positive for CD68 but not for myeloperoxidase⁵.

KFD usually follows a self-limited course with a possible recurrence rate of 3% to 4% reported in literature.³ Treatment is generally supportive since no specific treatment is available for Kikuchi's disease. Nonsteroidal anti-inflammatory drugs (NSAIDs) are used to alleviate the tender lymphadenitis and fever. Corticosteroids have been recommended if the patients manifest with severe extranodal or generalized disease, but this is of unproven efficacy⁶. Limited success has been reported with the use of intravenous immunoglobulin⁷

CONCLUSION

Kikuchi's disease must be considered in the differential diagnosis of cervical lymphadenopathy in a young female in order to avoid laborious investigations and potentially harmful treatments for infectious and lymphoproliferative diseases.

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