Case Report

Kikuchi Disease - A Rare Reality

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ABSTRACT

Kikuchi disease is a rare cause of cervical lymphadenopathy that is mostly seen in young women. This report describes the case of a 14 years old boy who presented with cervical lymphadenopathy. He had a history of enlarged cervical nodes about 15 days ago for which an excision biopsy was done that reported it as a case of Kikuchi’s disease. He was given a course of Non Steroidal Anti Inflammatory Drugs and the nodes subsided within 2 weeks. There are a very few reports of this disease in literature and few cases have been reported from India. Awareness regarding this entity is of crucial importance as Systemic Lupus Erythematosus (SLE) or lymphoma may present a similar picture on LN biopsy. Awareness of this disorder by clinicians and pathologists in India will help prevent misdiagnosis and inappropriate treatment.

Keywords: Kikuchi disease, Cervical Lymphadenopathy

INTRODUCTION

Kikuchi-Fujimoto disease or histiocytic necrotizing Lymphadenitis is a rare, benign, self limiting cervical lymphadenitis of unknown etiology (1,2) predominantly affecting young adults. It may be accompanied by mild fever and night-sweats. Prevalence among Japanese & other Asiatic individuals is higher (3) although it has a worldwide distribution. Presence of post cervical lymphadenopathy is an important characteristic feature in this condition. It is diagnosed by excisional lymph node biopsy. (4) Treatment is supportive with no definitive guidelines formulated, at least for time being.

CASE REPORT

A 14 year old male of Indian origin presented to us with swelling in multiple lymph nodes (LN) in neck and fever for 15 days prior to admission. No history of myalgia, arthralgia, night sweats, weight loss, bone pain. There was no history of any chronic drug intake. He did not suffer from tuberculosis previously or any contact with TB. Patient was a non smoker and non-alcoholic. There were no other medical problems. Similar problem was not noted among other family members. Clinical examination revealed that lymph nodes were palpable bilaterally in cervical region.- both submandibular, both jugulodigastic, right supraclavicular, left post auricular,
occipital lymphnodes; right submandibular one being the largest one(3.5×2.5cm²). All were soft, mobile, non-tender. Overlying skin was normal, no discharging sinus was noted. No lymph node was palpable in other part of the body. No sternal tenderness was noted. Blood pressure was 122/76 mm Hg. Pulse was 76/min, regular sinus rhythm. Mild pallor was present. Cardiovascular, respiratory, nervous system examination revealed no significant abnormality. Abdomen examination revealed mild hepatomegaly but no splenomegaly. Free fluid was absent. Bowel sounds were normal. Skin exam & throat exam were normal. Baseline investigations were within normal limit. Hemoglobin was 11.5g%, TLC-3400 N:\L; E:\B\M². Urea - 23mg%, Creatinine-0.7mg%, Bil-0.7mg%, TotalPr-6.49g%, Alb-3.69%, Glo-2.8g%, Alkaline phosphatase -115U/L, ALT-25U/L, AST-41U/L, Na⁺, K⁺-within normal limit. USG abdomen - few periportal LN were noted, Chest Xray was absolutely normal. Mantoux test was negative (induration 4 mm). Sputum AFB exam revealed no AFB. Excisional LN biopsy (scar mark shown in fig1) was done and histological picture suggested necrotizing histiocytic lymphadenitis compatible with the diagnosis of kikuchi disease (fig 2& 3). Work up for autoimmune disease was undertaken, Systemic Lupus Erythematosus exclusion work up was done. Antinuclear Antibody (ANA) and anti dsDNA came out to be negative. Bone marrow examination was within normal limits. In the meantime patient was put on oral NSAIDs. Fever and swelling in LN disappeared within two weeks.

**DISCUSSION**

Kikuchi first reported this disease in 1972. Fujimoto et.al independently described the disease entity in the same year. Cause of this disease is not known definitively but viral autoimmune mechanisms have been suggested. Females are more affected than males (F:M=3:1 - 4:1). Kikuchi’s disease mainly affects cervical LN and may be tender and is usually accompanied by fever
and Upper Respiratory Tract Infection. Arthralgia, myalgia may be complained by some patients. Weight loss, non specific chest pain, abdominal pain are also not uncommon. Physical exam may reveal hepatosplenomegaly. Exact etiology is not elucidated till now. Possible association with HIV, EBV, HTLV has been postulated. According to some studies, Toxoplasma, Bartonella may be causative organism. Frequent association with SLE has led to the probability of autoimmune mechanism being one of the causative factors although more research is necessary before such association is proved beyond doubt. It is suggested that CD8 cytotoxic T cells enter the LN and mediate apoptotic cell death which may form the necrosis in LN. Hypothetically, apoptotic lymphocytes could deliver nuclear antigens and trigger autoimmune T and B cells to produce antinuclear antibodies. Histocytes may act as enhancers. Role of IL6, IFN-gamma & IL2 have been noted in some cases. Routine lab measurements are usually normal in this disease although lymphocytic leucocytosis may be present. Some patient may have a diminished WBC count. ESR and C-Reactive Protein may be raised. Peripheral blood smear may reveal atypical lymphocytes. FNAC of LN is of limited diagnostic importance; one should not attach too much importance to it. Excisional LN biopsy is the investigation of choice. Morphologically it is characterised by irregular paracortical areas of necrosis with abundant karyorrhectic debris which may distort the nodal architecture. Large number of different types of histiocytes are present at the margin of necrotic foci. Karyorrhectic foci are formed by different cellular types, mainly histiocytes and plasmacytoid monocytes and also immunoblasts & lymphocytes. Neutrophils are characteristically absent. Immunophenotype of Kikuchi’s disease is composed of CD8(+) & CD4(+) T lymphocytes. KFD may be confused with SLE and malignant lymphoma. Almost total absence of plasma cells in biopsy differentiates it from SLE. Serological tests like ANA, anti dsDNA can be done in this setting. Again, incomplete architectural effacement, presence of patent sinuses, absence of Reed Sternberg cells, low mitotic rates renders it clear from lymphoma. No special treatment is available which is mainly supportive and NSAIDs being usual drug of choice. Corticosteroids may be indicated in severe cases. Role of intravenous immunoglobulin has not been clear but tried with limited success. The disease runs a benign course and is self-limiting in less than 4 weeks. Fatalities are rare and when occur, result from cardiac or respiratory failure. Recurrence rate is variable, around 3-4%. 

CONCLUSION

Although the disease is very rare, the clinician as well as the pathologist must be aware of the entity when a case of cervical lymphadenopathy in a young adult is being evaluated. Early recognition of this disease helps to minimize potentially harmful and unnecessary evaluations and treatment.

REFERENCES

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