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Case Report

Kikuchi Fujimoto disease: a rare case report

Sumit Sourabh[1]*, Akriti Jindal[1], Ashiana Singh[2]

[1]Department of Medicine, Chauhan hospital, Pathankot, Punjab, India
[2]Department of Paediatrics, Civil hospital, Gurdaspur, Punjab, India

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***Correspondence:**
Dr. Sumit Sourabh,
E-mail: drashiana1992@gmail.com

ABSTRACT

Kikuchi disease, also known as Kikuchi histiocytic necrotizing lymphadenitis, was initially described in young Japanese women. It is a rare benign self-limiting disease of unknown cause usually characterized by fever and cervical lunphadenitis. It is diagnosed by doing excisional biopsy of affected lymph node. A case of young 20 years old female presented with complaint of fever and weight loss for 20 days along with cervical and axillary lymphadenopathy. Complete physical examination, radiological investigations and biochemical tests were done to rule out systemic lupus erythematous, non-Hodgkin lymphoma and tuberculosis. Only symptomatic treatment was done along with corticosteroids. Its diagnosis is important as it can be easily mistaken for other form of lymphadenitis. Clinician and pathologists' awareness of this disorder is very necessary.

Keywords: Histiocytic necrotizing lymphadenitis, Kikuchi disease, Lymphadenitis

INTRODUCTION

Kikuchi disease is a benign self-limiting disease characterized by regional lymphadenopathy with tenderness, mild fever and night sweats. Other less common symptoms are weight loss, nausea, vomiting.[1] Its diagnosis is crucial as it can be easily misdiagnosed as tuberculosis, systemic lupus erythematosus or malignant lymphoma.[2] Most common group of lymph node involved is posterior cervical lymph nodes. The diagnostic modality is excisional biopsy showing fragmentation, necrosis and karyorrhexis.[1] Here we present a case of 20 years old female patient with fever, painful cervical and axillary lymphadenopathy and weight loss. This also discuss practical steps for diagnosing and managing Kikuchi disease.

CASE REPORT

A 20 years old female admitted to our hospital with complaint of fever max. documented to 101 F, weight loss for 20 days. On physical examination, two swellings were observed in right neck with maximum diameter being about 2 cm and were soft and tender on palpation. A single tender axillary lymph node was also observed in left axilla. With patient's consent, medical examination and investigations were done. Complete blood count was normal, CRP was 10.8 mg/l and ESR was 45mm. The results from chest X-ray as shown in Figure 1 and tuberculin test were negative ruling out tuberculosis.

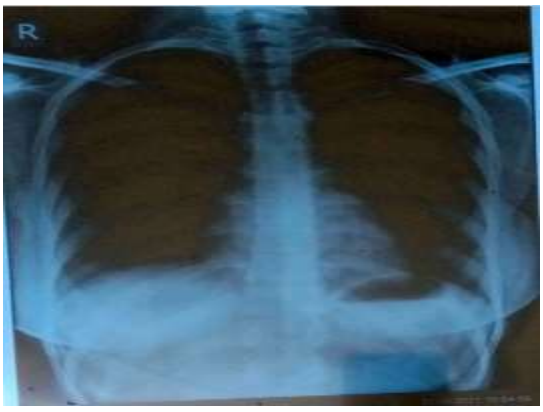

Figure 1: No active sign of tuberculosis.

USG guided FNAC was done showing hypercellular smear with population of lymphoid cells consisting of lymphocytes, centrocytes and centroblasts with lymphohistiocytic aggregates as well. Histopathological examination was further suggestive of histiocytic necrotizing lymphadenitis as shown in Figure 2 (A and B).

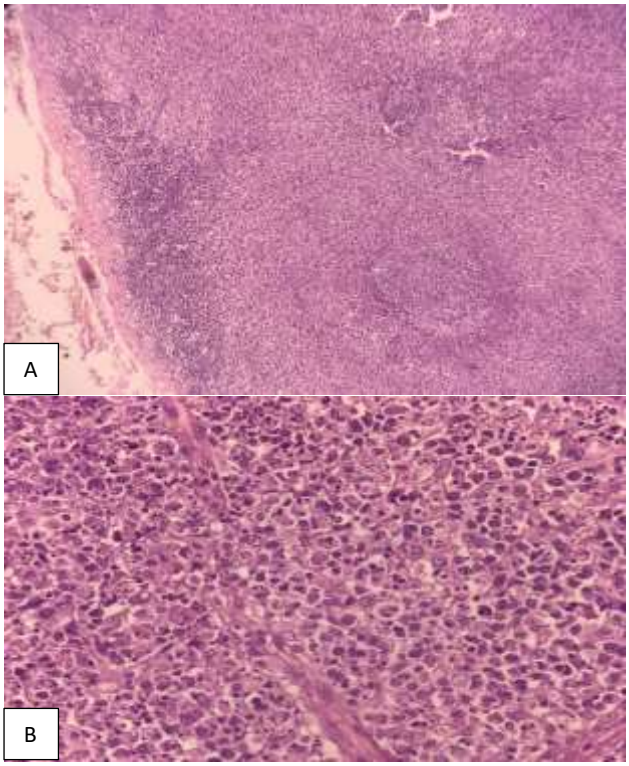

Figure 2 (A and B): Focal, paracortical necrotizing lesion (H and E 100 X), mononuclear infiltrate along with karyorrhexis/pyknosis, nuclear debris and scattered fibrin deposits (H and E 400 X).

Further investigations were done to rule out SLE. Lab studies were negative for hepatitis B, hepatitis C, HIV, VDRL. Anti dsDNA was within normal limit (11.02IU/ml) while ANA was negative (11.7 units). Initially the patient was started on NSAIDS (aceclofenac) but patient shows no improvement, so methylprednisolone at 1mg/kg was started. She was given this dose for 7 days and then this dose was tapered in every consecutive three days. The patient was scheduled for close follow up and discharged. At next follow-up in one month, lymphocytosis had regressed, CRP and ESR return to normal level and size of lymph nodes were decreased. Two months later, physical examination and laboratory measurements were within normal limits and methylprednisolone was discontinued. The patient is not receiving any treatment after 3 months of follow up.

DISCUSSION

KFD is a disease which presents as fever, sweats and progressive painful lymphadenopathy mainly involving cervical regions. A viral or autoimmune cause has been found out. Another theory says that KFD may represent an exaggerated T cell mediated immune response in genetically susceptible individual to a number of stimuli. This disease is first reported in japan.[3] Most common age group is 30-40 years, with female to male ratio 4:1.[1]

Lymph node size has been found to range from 0.5 to 4 cm, but it may reach 5 to 6 cm and rarely larger than 6 cm. Lymphadenopathy is rare.[4] 50% patient with KFD may have fever. Fever is usually of low grade and mostly associated with upper respiratory symptoms. Others symptoms include weight loss, nausea, vomiting, sore throat and night sweats. Leucopenia is seen in 50% of cases. Atypical lymphocyctes in the peripheral blood can be seen. Involvement of extra-nodal sites in KFD is uncommon.[3]

The diagnosis depends on excisional biopsy with characteristic histopathologic findings of KFD, including paracortical areas of coagulative necrosis with abundant karyorrhectic debris, distortion of the nodal architecture, and large number of histiocytes at the margins of necrotic areas.[1]

Differential diagnosis of lymph node enlargement includes tuberculosis, SLE, lymphoma. The histological differential diagnosis of KFD mainly includes lymphadenitis associated with SLE, herpes simplex, non-Hodgkin lymphoma, plasmocytoid T cell leukemia, Kawasaki disease, myeloid tumor.[3] Differentiation of KFD from SLE is done on the basis of C3, C4, anti-Sm and LE cells. KFD may be misdiagnosed as malignant lymphoma. Features of KFD that differentiate it from malignant lymphoma are incomplete architectural effacement with patent sinuses, presence of numerous reactive histiocytes, absence of Reed-Sternderg cells.[5]

The process is typically self-limited showing recovery from one to six months, 3-4% cases may show reocurrance.[1,6] Lymphadenopathy can resolve spontaneously. Treatment includes analgesia, antipyretics and NSAIDS therapy. Corticosteroid treatment is recommended if extra-cervical manifestations are present.[1] Therapeutic responses to minocycline and ciprofloxacin have been used as supportive evidence for an infective aetiology in Kikuchi's disease.[7,8] Apart from this, two case reports describe a therapeutic response to combined treatment with IVIG and corticosteroids in children with haemophagocytic syndrome with Kikuchi 's lymphadenitis. No risk to other family members is felt to be associated with KFD.[5]

CONCLUSION

Currently no vaccine is available for its prevention. The relationship between this disease and infections still not well established. Reduction in fever and malaise is the first sign of improvement followed by reduction in size of lymph nodes. As Kikuchi is a very rare disease, several questions need further investigations.

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