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KIKUCHI-FUJIMOTO DISEASE: A RARE CAUSE OF LYMPHADENOPATHY

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ABSTRACT
Kikuchi-Fujimoto disease or Necrotising Histiocytic Lymphadenopathy is a rare condition presenting as lymphadenopathy and occasionally with fever. We present a case of a 22 year old lady who presented with fever and cervical lymphadenopathy. The diagnosis was made after a lymph node biopsy. The patient was managed conservatively and complete recovery was seen on subsequent followup. In a country like India where Tuberculosis is rampant, It is essential to keep in mind the possibility of rare diseases to avoid the cost of unnecessary treatment and distress to the patient.

KEYWORDS: Necrotising Histiocytic Lymphadenopathy, cervical lymphadenopathy.

BACKGROUND

A patient presenting with fever and lymphadenopathy leads to prompt investigations for tuberculosis and lymphoma. However, rare conditions like kikuchi-Fujomoto disease must be kept in mind and a lymph node biopsy followed by histopathologic examination should be done to confirm the diagnosis and avoid the cost of unnecessary treatment and distress to the patient.

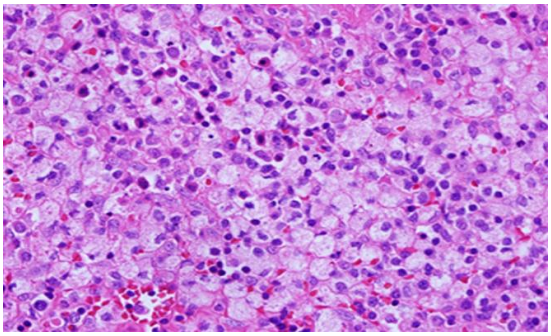
CASE PRESENTATION

A 22 year lady presented with the complaints of on and off fever for 2 months associated with fatigue. The patient was previously fit without any history of any chronic disease or any ongoing long term treatment. On examination the patient was febrile and hemodynamically stable. There were palpable cervical lymphnodes bilaterally, which were firm and non-tender with a maximum size of 2 cm x 3 cm. There was no palpable hepatomegaly or splenomegaly. Blood investigations revealed-

PARAMETER	Patient value	NORMAL RANGE
Hb	11.3 gm/dl	11.0-15.0
TLC	3810 /µl	4000-10000
DLC (N/L/E/M)	46/45/7/2	
PLATELET	175000/µl	150000-450000
MCV	84 fl	80-100
LDH	376 U/L	230-460
Renal profile		
CREATININE	0.7 mg/dl	0.5-1.4/15-45
UREA	23 mg/dl	15-45 mg/dl
SODIUM	140 mmol/l	135-145
POTASSIUM	3.6 mmol/l	3.5-5.5

Liver profile		
SGOT / SGPT	28/13 U/L	9-39/10-40
TOTAL BIL./DIRECT BIL.	0.5/0.2 mg/dl	0.1-1.2/0.01-0.3
TOTAL PROTEIN/ ALBUMIN	7.8/4.4 g/dl	6.4-8.5/3.2-5.5
ALKALINE PHOSPHATASE	107 U/L	110-310
Autoimmune panel		
ANA	0.2	<1.2
Anti dsDNA	20 IU/ml	<30

- ECG and Chest X-Ray were within normal limits.
- USG Abdomen : Normal study
- HIV / HBsAg /Anti-HCV : Non Reactive
- FNAC from cervical lymph node : Reactive Lymphoid Hyperplasia
- Histopathology from cervical lymph node biopsy:



Histopathology of lymphnode biopsy -Features strongly suggestive of KIKUCHI'S disease.

The patient was managed conservatively with paracetamol. On follow up, the patient improved and lymphadenopathy subsided.

DISCUSSION

KFD is a benign histiocytic necrotising lymphadenitis. Its aetiology is still unclear, however viral origin, EBV, HHV6 and 8 have been postulated.[4] An autoimmune aetiology is also likely as it has been reported in association with SLE. It tends to affect a young population under 30 years of age, including children, although the latter are less commonly affected. Prevalence is found to be almost equal in both genders.

The most common clinical manifestations include lymphadenopathy, fever, sweats, malaise, anorexia, weight loss, hepatomegaly and leucopenia.[1]

A diagnosis is made by lymphnode biopsy. Histopathological assessment of affected lymph nodes reveals characteristic findings. There are three main patterns identified, proliferative, necrotizing and xanthomatous. The proliferative picture is seen in approximately a third of cases and has a dominant inflammatory infiltrate. Half of cases show necrotizing pattern and the xanthomatous type is rare and has abundant foam cells.[2] As the symptoms are non-specific and some of the histological features are similar to other diseases it is easy to misdiagnose KFD with SLE or lymphoma. This is important as the treatment of KFD is symptomatic and supportive, spontaneous recovery is usual, while the latter two conditions require prompt specific treatments.

Long term follow-up of these patients is necessary as recurrent cases of KFD have been reported and there is some belief that KFD may be a precursor for SLE.

CONCLUSION

Kikuchi-fujimoto disease is rare but should be kept in mind as a differential diagnosis of cervical lymphadenopathy. Identification is important because it is essentially self limiting and thus varies considerably from other disorders which have a similar clinical presentation like tuberculosis, Lymphoma and SLE. The diagnosis is confirmed by lymphnode biopsy. The patient should be kept on long term followup so as to ascertain resolution, detect recurrence or development of SLE.

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