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A case of Kikuchi disease presenting with aseptic meningitis

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I. Introduction

Histiocytic necrotizing lymphadenitis (HNL) is a benign and self-limiting systemic disorder, involving lymph nodes with clinical manifestations of mild fever, superficial lymphadenopathy and rash among others. It is commonly known as Kikuchi-Fujimoto disease (KFD) as first reported in Japan by Kikuchi [1] and Fujimoto [2] in 1972.

Kikuchi-Fujimoto Disease typically follows a benign and self-limited course, characterized by cervical lymphadenopathy (most common). Less frequently, other symptoms might also be present like nausea, weight loss, night sweats and fatigue (3). It is often misdiagnosed given its overlapping clinical presentation with lupus and lymphoma. Generally, KFD is diagnosed via excisional lymph node biopsy and histopathological analysis. KFD shares many characteristics with other causes of lymphadenopathy including lymphoma, inflammatory disorders, autoimmune conditions, and infectious causes of lymphadenopathy like tuberculosis infection; therefore, it is important consider KFD in cases of persistent lymphadenopathy and must be differentiated from these conditions (3). Kikuchi-Fujimoto disease constituted 2.1% of 1724 lymph node biopsy specimens in a study published from Tirupati[4] and has been occasionally reported from India. We report this case of Kikuchi-Fujimoto disease with CNS involvement in the form of meningitis.

II. Case report

We report a case of 20 years old female presented with complaints of fever since 4 days along with altered sensorium since 1 day. Patient had history of headache and vomiting since last 3 days followed by altered sensorium .

PAST HISTORY -She also had history of intermittent fever since last 2 months . Fever was associated with arthralgia (on and off) for same duration ,one episode of which resulted in hospitalization 2 weeks back .Her previous medical reports suggested her mean temperature during earlier admission to be 37.9°C,Blood parameters suggested neutropenia ,raised ESR of 42 mm/hr and raised CRP .Patient responded to antipyretics and NSAIDs and was discharged on day 3 .

ON PHYSICAL EXAMINATION

- BP -130/80 mmHg
- PR -122 beats/min regular in rhythm normal in volume and character .
- Neck rigidity – present .
- Kernig sign –present
- A palpable tender lymph node approx. (2*1.5)cm was present in supraclavicular fossa .
- Multiple palpable lymph nodes were present in cervical region .

Her baseline blood parameters were

Parameter	Patient value	Normal range
Hb	11.7 gm/dL	11.0-15.0
TLC	8000 cells/mm^3	4000-11000
Neutrophils	69%	40-75
Lymphocytes	24%	20-50

Eosinophils	4%	1-6
Monocytes	2%	2-10
Basophils	1%	<1
Platelet count	150,000 cells/mm^3	150,000-350,000
RBC count	4.1 x 10^6 cells/mm^3	4-6
ESR	72	<15
CRP	9.2 mg/dL	<0.3
S.Creatinine	0.6 mg/dl	0.2-1.5 mg/dl
SGOT	14 mg/dl	Upto 37 mg/dl
SGPT	22 mg/dl	Upto 42 mg/dl
T.bilirubin	0.7 mg/dl	0.3-1.2 mg/dl
Direct bilirubin	0.4 mg/dl	0-0.35mg/dl
Indirect bilirubin	0.2 mg/dl	0.2-0.65 mg/dl
LDH	256 U/L	230-460

AUTOIMMUNE PANEL
ANA by IFA - negative

CSF ANALYSIS

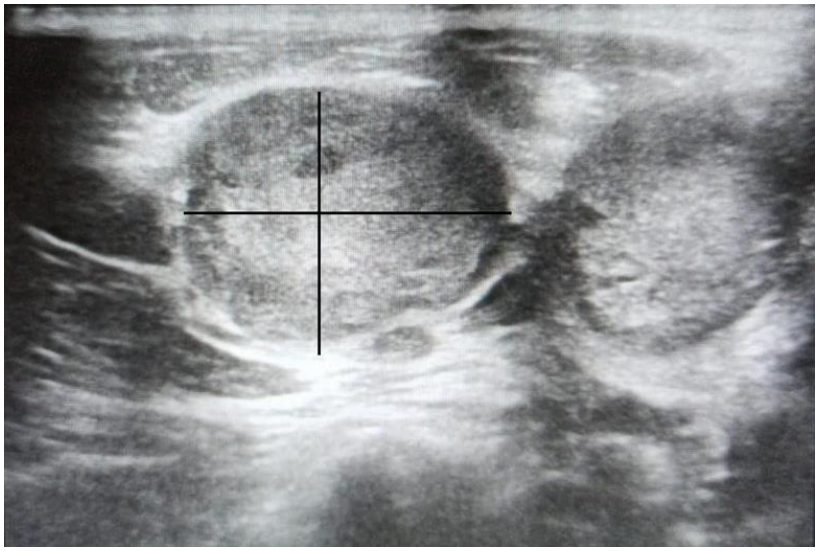
Cell count	4 cells (all lymphocytes)	<5
Protein	30 mg/dl	15-45 mg/dl
Glucose	73 mg/dl	50-80 mg/dl
ADA	07 MG/DL	<10 mg/dl

CSF for Acid Fast Bacillus –negative. CSF culture – no microbial growth

MICROBIOLOGICAL PANEL
PBS for malarial parasite – negative Typhidot - negative Dengue NS1 ANTIGEN – negative IgM for Scrub Typhus – negative IgM for Leptospira- negative

RADIOLOGICAL ANALYSIS

USG neck showed multiple prominent to enlarged discrete lymph nodes in **right cervical regions at level III, IV, V and right supraclavicular fossa without any evidence of calcification and necrosis.**Non Contrast CT BRAIN didn't reveal any significant parenchymal abnormality .Plain Chest skiagram didn't reveal any abnormality.



USG neck showing enlarged lymph nodes with loss of hila .largest node is approx 25*13mm

HISTOPATHOLOGICAL AND IMMUNOHISTOCHEMISTRY EXAMINATION

Excisional biopsy was done from level 5 cervical lymph node and sections were made.Sections show partial effacement of lymph node architecture along with irregular necrotic areas with large number of apoptotic body and histiocytes .The lymph node biopsy disclosed histopathologic features typical of histiocytic necrotizing

lymphadenitis(Kikuchi fujimoto disease) with CSF picture suggestive of aseptic meningitis

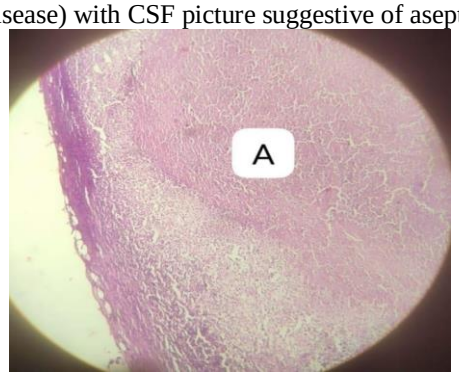


Image 2 :Lymph Node Capsule With Areas Of Necrosis And Distortion Of Architexture

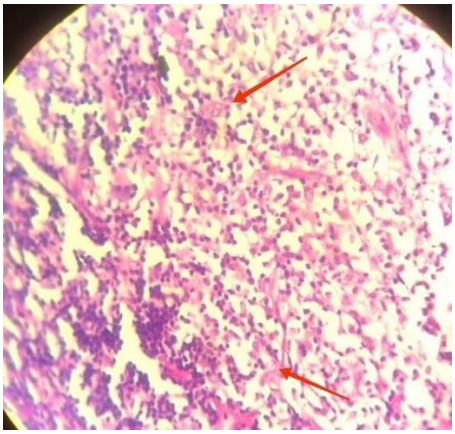
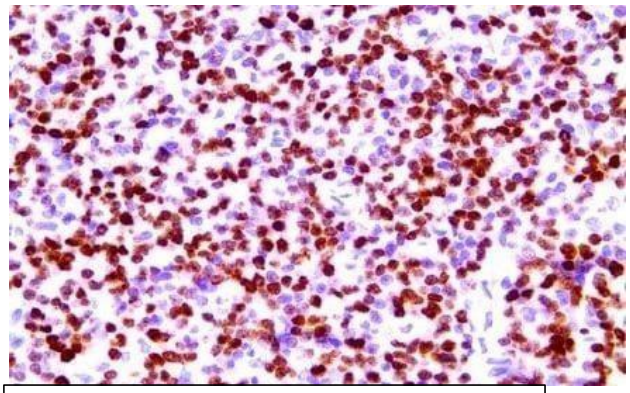


Image 3 :Large Number Of Histiocytes In Background Of Necrosis.No Granuloma Seen



Ki67 staining of lymph node biopsy specimen

Management

Patient was started on intravenous steroids (dexamethasone 8mg)three times daily which was gradually tapered along with antibiotics and antivirals. Patient responded to steroids and sensorium improved and was discharged on DAY 10 .She was given NSAIDs on discharge for her tender nodes along with oral steroids which was gradually tapered over 1 month and was followed up.

Followup

Patient was followed up and after 4 weeks lymphadenopathy started decreasing .No complaints of fever was found during followup.Lymphadenopathy completely disappeared clinically at the end of 12 weeks .

III. DISCUSSION

Kikuchi disease is a rare cause of cervical lymphadenitis. It is a benign,self-limiting disease; however, it is frequently misdiagnosed, and patients are subjected to unnecessary treatments. One study found that Kikuchi

disease was misdiagnosed in 40% of cases, most often diagnosed as lymphoma(5)

The exact incidence of kikuchi disease is unknown but it was found to be 5.7% in cases presented with lymphadenopathy. It most commonly affects young females with a mean age of 21 years and a female predominance of 2:1, although it can occur in males and older individuals. The disease is typically found in Japanese individuals and other Asian populations; however, it has been described in all races.

The clinical course of this disease has some specific and non-specific features with the specific one being unilateral cervical lymphadenopathy. (6) .Most common KFD manifestations include fever, weight loss, sweats, lymphadenopathy, anorexia, hepatomegaly, and lymphopenia. Several differential diagnoses are associated with this presentation (7). The clinical presentation with a prodrome of sinopulmonary infection, lymphadenopathy, atypical lymphocytosis, and lack of antibiotic response strongly suggest the postulated viral etiology, even though the etiology remains largely unclear. The electron microscopic features suggestive of reticular tubular structures inside the cytoplasm of activated histiocytes and lymphocytes, which are found in KFD, have also been observed in other autoimmune disorders such as SLE, thus pointing to an autoimmune process as the possible etiology as well (6) The most common laboratory abnormalities are a mild leukopenia seen in 50% of cases and elevated inflammatory markers with an erythrocyte sedimentation rate >60 .Infection with unknown pathogens can produce causative substances which may bind to different parts of CNS .

KFD is a rare disorder which should be considered in differential diagnosis of patients presented with fever and lymphadenopathy .Patients with KFD can develop neurological symptoms whose spectrum can be as vivid as aseptic meningitis,multiple neuritis or acute cerebellar ataxia.Conditions such as tuberculosis meningitis ,SLE and infections should be excluded and the possibility of encephalopathy associated KFD should be considered.

Management includes glucocorticoids along with immunoglobulins if not responded. Antiviral therapy should be added if viral infections cannot be ruled out completely.Close follow up is emphasized in these patients(12).

IV. Conclusion

Although a rare cause Kikuchi disease should be considered in differential diagnosis of patients of altered sensorium along with lymphadenopathy .Biopsy should be performed early to avoid misdiagnosis and unnecessary treatment.Long term followup of these patients are required to monitor for disease recurrence .

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