International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Kikuchi Fujimoto Disease - A Rare Case

Dr. Tuhin Shrivastava[1], Dr. Urvee Swaika[2], Dr. Rupali Jain[3], Dr. Urshlla Kaul[4], Dr. Vijay Vishwanathan[5], Dr. Vijay Kamale[6]

Abstract: ***Patient was a 14 year old male came with complaints of high grade fever and chills since 2 weeks, along with swelling in the left axillary area with tenderness. There was history of extrapulmonary tuberculosis in grandmother 2 years ago for which adequate treatment was taken. His CRP was 3.9 (normal <0.5), ESR 38 and total leucocyte count of 4410. Management included sending workup in the lines of 'pyrexia of unknown origin'. Investigations done for Dengue, malaria WIDAL, scrub typhus, EBV, brucella, leptospira and atypical pneumonia, auto immune workup all of which were negative. Ultrasound of left axilla and HRCT chest were suggestive of multiple enlarged non necrotic lymph nodes left axillary and pectoral plane. CECT abdomen, gastric lavage for AFB and CBNAAT done to rule out tuberculosis which were negative. In view of pyrexia of unknown origin doxycycline, azithromycin and chloroquine trial given but there was no response to fever. Finally lymph node biopsy was taken in which lymph 3 enlarged lobulated lymph nodes excised and were sent for TB CBNAAT, MGIT, gram staining and histopathology out of which histopathology was suggestive of Kikuchi Fujimoto disease. Post lymph node excision fever subsided immediately. Patient was started on oral naproxen. Hence it took a long battle to differentiate between uncommon infectious causes, tuberculosis and autoimmune causes finally to end with a rare diagnosis of Kikuchi Fujimoto disease which is usually uncommon at this age and gender.***

Keywords: kikuchi fujimoto, lymph node, histiocytic necrotizing lymphadenitis, rare disease, pyrexia of unknown origin, lymph node biopsy

1. Introduction

Kikuchi-Fujimoto disease (KFD) is an enigmatic, benign and self-limited syndrome characterized by regional lymphadenopathy with tenderness, predominantly in the cervical region, usually accompanied by mild fever and night sweats. Initially described in Japan, KFD was first reported in 1972 almost simultaneously by Kikuchi and by Fujimoto *et al*. It is also known as histiocytic necrotizing lymphadenitis.

Less frequent symptoms include weight loss, nausea, vomiting, sore throat. Kikuchi-Fujimoto disease is an extremely rare disease with only approximately a few hundred reported cases. The clinical, histopathological and immunohistochemical features appear to point to a viral etiology, a hypothesis that still has not been proven. KFD is generally diagnosed on the basis of an excisional biopsy of affected lymph nodes.

2. Case Report

14 year old male came with complaints of high grade fever and chills since 2 weeks, along with swelling in the left axillary area with tenderness. There was history of extrapulmonary tuberculosis in grandmother 2 years ago. All baseline investigations were normal with normal total leucocyte counts and normal CRP. ESR was raised. Management included sending workup in the lines of 'pyrexia of unknown origin', all of which were negative. Ultrasound of left axilla and HRCT chest were suggestive of multiple enlarged non necrotic lymph nodes left axillary and pectoral plane. Tuberculosis and SLE was ruled out.

There was no response to antibiotics. In view of pyrexia of unknown origin doxycycline, azithromycin and chloroquine trial given but high grade fever still persisted. Finally lymph node biopsy was taken in which lymph 3 enlarged lobulated lymph nodes excised and were sent for TB CBNAAT, MGIT, gram staining and histopathology out of which histopathology was suggestive of Kikuchi Fujimoto disease. Post lymph node excision fever subsided immediately. Patient was started on oral naproxen. for 4 weeks and asked to follow up.

3. Discussion

KFD is generally diagnosed on the basis of an excisional biopsy of affected lymph nodes.

Histopathological examination showed abundant necrotic areas with numerous histiocytes and extensive nuclear debris and focal proliferation of reticular cells and neutrophils are absent–which is important for diagnosis.

There is focal proliferation of reticular cells accompanied by numerous histiocytes and extensive nuclear debrisProliferating CD8 positive T-cells may act as "killers" and "victims" in the apoptotic process *via* fas-and perforine-pathways. A low but possible recurrence rate of 3 to 4% has been reported. In few patients, SLE may occur some years later.

4. Conclusion

Its recognition is crucial especially because this disease can be mistaken for systemic lupus erythematosus, malignant lymphoma or even, though rarely, for adenocarcinoma.

The differential diagnosis of fever and cervical lymphadenopathy is broad and often leads to an extensive workup.

Analgesics-antipyretics and nonsteroidal anti-inflammatory drugs may be used to alleviate lymph node tenderness and fever. Surgical consultation may be indicated for a diagnostic excisional lymph node biopsy.

Spontaneous recovery occurs in 1 to 4 months. Patients with Kikuchi-Fujimoto disease should be followed-up for several years to survey the possibility of the development of systemic lupus erythematosus.

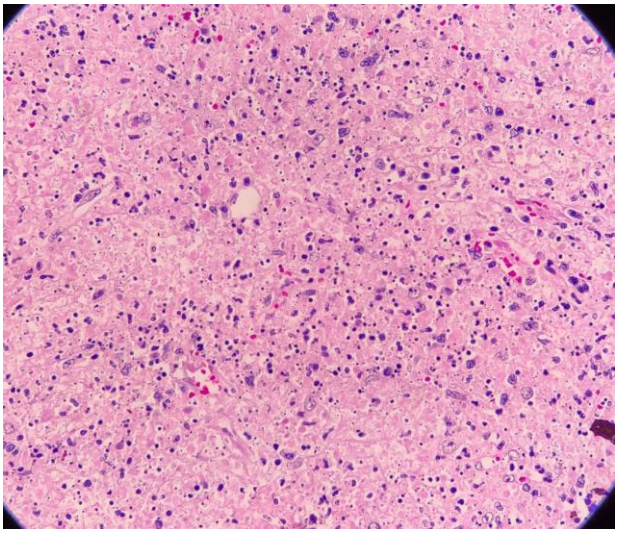

Figure A: Pale necrotic areas with karyorrhectic debris (nuclear dust)

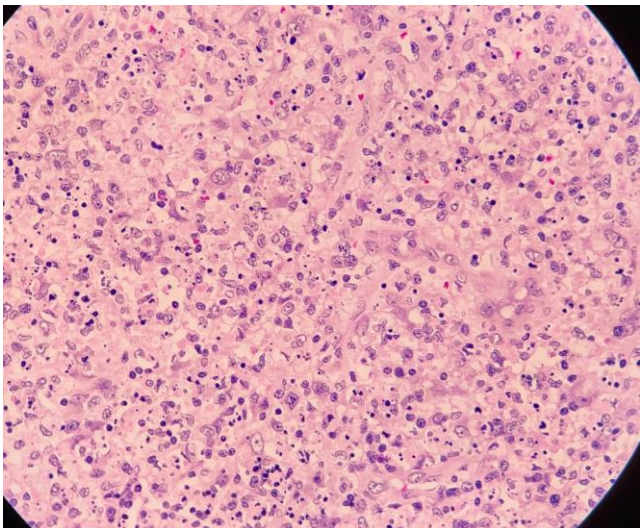

Figure B: Histiocytes with few lymphocytes and karyorrhectic debris. Absence of neutrophils

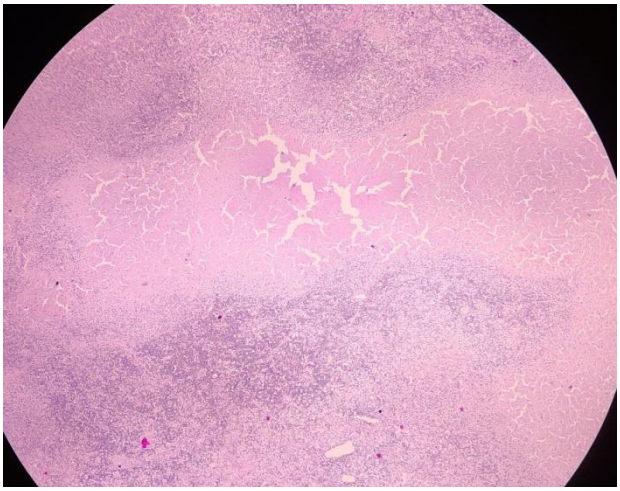

Figure C: Lymph node capsule with distorted lymph node architecture

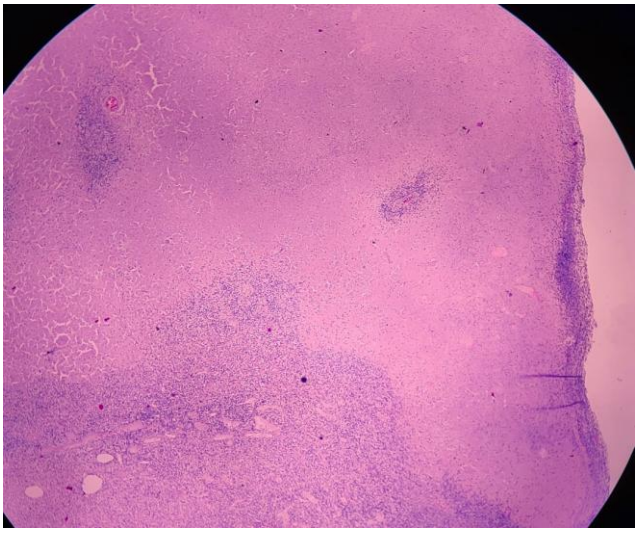

Figure D: Serpiginous pale stained areas of necrosis