

**A Case of Kikuchi-Fujimoto Disease in an Indian Female and its
Management: A Case Report**

Abstract

Kikuchi-Fujimoto disease is a self-limiting illness characterised by non-specific symptoms, most commonly reported symptoms being fever and lymphadenopathy. The etiological cause of this disease remains unknown. This condition needs to be differentiated from other closely resembling conditions such as tuberculosis, lymphoma, Systemic Lupus Erythematosus (SLE) and adenocarcinoma. The physician needs to be aware about this condition to avoid misdiagnosis and improper treatment.

Keywords

Kikuchi-Fujimoto syndrome, Benign, Lymphoma, Tuberculosis, Lymphadenopathy

Introduction

Kikuchi-Fujimoto disease also known as histiocytic necrotizing lymphadenitis, Kikuchi necrotizing lymphadenitis, phagocytic necrotizing lymphadenitis, subacute necrotizing lymphadenitis, and necrotizing lymphadenitis, was first described in 1972 in Japan by Dr Masahiro Kikuchi and by Y.Fujimoto independently. ^[1-5] Kikuchi-Fujimoto disease (KFD) is a rare, self-limiting disorder that typically affects the cervical lymph nodes. ^[1] The presentation of Kikuchi-Fujimoto disease is variable, and there is no specific set of symptoms or laboratory features that reliably establishes the diagnosis. ^[6] Kikuchi disease occurs sporadically in people with no family history of the condition. ^[5] Very few cases of Kikuchi-Fujimoto disease have been described in Indian patients. It is especially important to differentiate this condition from tuberculosis because of the high prevalence of tuberculosis in Indian population, close resemblance and different treatments. Other close differential diagnosis includes lymphoma and adenocarcinoma ^[1]. A physician needs to be aware of this condition to prevent misdiagnosis and inappropriate treatment. We hereby describe a case of Kikuchi-Fujimoto disease in a young adult female of Indian descent.

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Case Presentation

A 26-year-old Indian female who had no major prior medical history arrived at our outpatient department with complaints of fever and neck swelling that developed over a course of 30 days. She was treated at a local hospital with empirical antibiotics with no improvement of symptoms. No drug or alcohol use was reported. Family history was unremarkable. She was a home-maker from Madhya Pradesh and had been married for 4 years. Initially, she was given treatment for seven days at a local hospital with no improvement in symptoms. She was referred to our hospital for additional treatment due to her ongoing fever and bilateral cervical region swelling. Indoor admission consent was obtained from the patient and attendants. On admission to the female medical ward, the patient was noted to be febrile. She also had bilateral cervical lymphadenopathy which ranged from 0.8-1.2 cm in size with no complaints of cough or breathlessness. On admission, her heart rate was 128/min, her respiratory rate was 20/min, her blood pressure was 120/70 mmHg, and her temperature was 40.2°C. Investigations performed on admission are mentioned in the following table.

Table 1- Investigations on admission

Laboratory Data	Results	Reference value
Haemoglobin	11.7	13-17
WBC	11,600	4000-11000
Platelet	2.4	1.5-4.5
ESR	20	<18
Urea	32	8-52
Serum Creatinine	0.8	0.6-1.3
RBS	74	70-126
HIV/HCV/HBSAG (Rapid tests)	Negative	
Sodium/Potassium	134/4.4	135-145/3.5-5.5
T. Bilirubin	0.6	0.2-1.0
CRP	28.4	0-6.0
SGOT	24	Up to 40
SGPT	24	Up to 40
Albumin	2.6	3.4-5.4
EBV mono spot test	Negative	
HIV (ELISA)	Negative	
P.S. For Immature or Atypical cells	Negative	
ENT examination	Normal	
H1N1 RTPCR	Negative	
COVID-19 RTPCR	Negative	
ANA Profile	Negative	
RA factor	Negative	
Anti-CCP Antibodies	Negative	
Urine Routine	Micro Albuminuria absent, Sugar absent, no cast	
Lymph Node Biopsy	Focal Areas of Non-suppurative Necrosis with Histiocytic and Plasmacytoid cell infiltrates	

A pulmonary and ENT (ear, nose and throat) examination was normal. Lab work revealed mild leucocytosis, elevated acute phase reactants and hypoalbuminemia with normal aspartate aminotransferase, alanine aminotransferase and platelet counts. Peripheral smear came out negative for atypical or immature cells. The serology for HIV, HCV and HbSAg was negative. The EBV mono-spot test and indirect fluorescent antibody test both were negative. Both the COVID-19 and H₁N₁ RTPCR results were negative. The samples of blood and sputum were sent for sensitivity and culture testing. The patient was given standard treatment. Investigational blood was sent, and blood for blood cultures was obtained. A lymph node biopsy was planned on the 4th day of admission and after meeting all pre-procedural requirements a lymph node biopsy was performed on the 5th day of admission.

The biopsy showed focal areas of non-suppurative necrosis with histiocytic and plasmacytoid cell infiltrates. The lymph node biopsy was negative for AFB (Acid fast bacilli) stain, atypical cells, immature cells and Gram's stain. Based on biopsy results, negative EBV serology and peripheral smear results a diagnosis of Kikuchi-Fujimoto syndrome was suspected.

She was managed symptomatically with anti-pyretics and NSAIDs for the pain. Fever resolved on the 7th day of admission and she was discharged on similar oral treatment. On follow up after 15 days, her fever had resolved with reduction in the size of lymph nodes. On further follow up after 2 months the lymphadenopathy had completely resolved with no other complaints.

Discussion

Kikuchi-Fujimoto disease (KFD), was first observed in Japan in 1972. Since then, it has been reported in a number of nations. It is widely acknowledged that KFD is a form of lymphoid reactive hyperplasia that causes non-neoplastic lymph node hypertrophy.^[7] KFD was formerly believed to be more common among women, however current research suggest the condition may affect both sexes.^[8]

Fever and cervical lymphadenopathy have a wide range of potential differential diagnoses, which frequently results in a thorough workup. Our patient was tested for lymphoma, tuberculosis and adenocarcinoma. The diagnosis was aided by the results of the lymph node biopsy. Necrosis and a histiocytic cellular infiltration can be found in one or more locations within the lymph node, which is the hallmark histology of KFD. There is a chance that the node's capsule will be penetrated, and perinodal inflammation is usual.^[9]

Even though autoimmune and infectious aetiologies have been put out as potential causes, KFD illness still has no established aetiology. There have been some suggested causal organisms. These include Brucella, Yersinia enterocolitica, Epstein-Barr virus, human T-cell leukaemia virus type 1, human herpesvirus type 6, B19 parvovirus, cytomegalovirus, and parainfluenza virus.^[10] Case reports include KFD developing concurrently with or after circumstances that may have acted as triggers, such as vaccination and COVID-19 infection.^{[11][12][13][14][15][16]}

Unilateral cervical lymphadenopathy, with or without systemic signs and symptoms like fever and skin rash, is the most typical clinical symptom of Kikuchi illness.^{[4][17][18][19]} In this case, our patient had bilateral cervical lymphadenopathy and fever.

Almost typically, Kikuchi disease has a benign course and clears up in a few weeks to months. For KFD, there is no proven effective treatment. It's a benign, self-limiting condition that goes away in one to four months. However, patients should be followed up on because they might later develop Systemic Lupus Erythematosus (SLE) or in rare cases, relapse with KFD. The latter does not frequently occur again.^[20] KFD is often treated with supportive care. Symptomatic relief is the main goal of treatment, which includes using analgesics and antipyretics to reduce fever and sensitive lymph nodes. For severe cases or recurring illnesses, corticosteroids are only used. It is advised that patients have a comprehensive survey and follow-up to assess any subsequent development of SLE because of the relationship with the disease. For steroid-resistant or recurring instances – hydroxychloroquine is indicated. Intravenous immunoglobulin is indicated for situations that are recurrent or resistant to steroids. Our patient was managed symptomatically with anti-pyretics and NSAIDs for the pain. Fever resolved on the 7th day of admission and she was discharged on similar oral treatment. On follow up after 15 days, her fever had resolved with reduction in the size of lymph nodes. On further follow up after 2 months the lymphadenopathy had completely resolved with no other complaints.

Why This Case Report was Important?

It is particularly important to distinguish this disorder from tuberculosis due to the high prevalence of tuberculosis in the Indian population, similarity, and distinct therapies. Other near differential diagnosis include lymphoma and adenocarcinoma^[1]. To avoid an incorrect diagnosis and the wrong kind of treatment, a doctor needs to be aware of

this illness. In our case we present a young adult female of Indian heritage who had Kikuchi-Fujimoto Disease.

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