



## A Rare Necrotizing Lymphadenitis: Kikuchi-Fujimoto Disease

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### ABSTRACT

Kikuchi-Fujimoto disease also known as necrotizing lymphadenitis is a rare, self limiting cause of lymphadenitis with unknown etiology and pathogenesis. The clinical signs and symptoms include cervical lymphadenopathy, fever, less frequent chills, night sweats, arthralgia, rash, weight loss and symptoms of respiratory infections. Diagnosis is often based on histopathological studies involving lymph node. It is often misdiagnosed with SLE, Lymphoma etc. There is no treatment guidelines given for KFD after diagnosis it is often limited to treat the symptoms but in severe cases corticosteroids can be given. Here we discuss a 14yr old female patient presented with evening rise of temperature and significant weight loss of approximately 5kg. Along with that she has also complained about bilateral axillary swelling of small size with pain. The final diagnosis was based on lymph node biopsy indicating Kikuchi-Fujimoto disease also the laboratory values were also elevated. After diagnosis patient was treated with symptomatically and the condition of the patient also improved. Although Kikuchi-Fujimoto disease can be misdiagnosed with other diseases, a systematic diagnostic approach is needed. Since there is no specific guidelines available for the management of Kikuchi-Fujimoto disease, further studies are needed to conduct the therapeutic option and its long term outcome.

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### INTRODUCTION

Kikuchi-Fujimoto disease (KFD) a rare, idiopathic and self limiting cause of lymphadenitis. It is also known as histiocytic necrotizing lymphadenitis (Lelii *et al.*, 2018). The etiology of KFD is

unknown but theories suggest that it is caused by one or more unidentified agents. KFD is often misdiagnosed as malignant lymphoma because of its acute and sub acute onset of adenopathy and systemic B symptoms and also associated with systemic lupus erythematosus (SLE) (Deaver *et al.*, 2014). It can mimic different pathologic conditions such as mononucleosis, lymphoma, metastases, and tuberculous lymphadenitis (Humphreys *et al.*, 2018; Kavirayani and Balan, 2017). The pathogenesis is unknown. The common symptoms of KFD include fever, cervical lymphadenopathy and elevated inflammatory markers. Increased ferritin level is indicated as one of the marker for diagnosis but it can also be due to certain other disease condition like co-occurrence of adult-onset still's disease (AOSD), or complication of reactive hemophagocytic lymphohistiocytosis. The main diagnosis marker is lymph node biopsy (Dalugama and Gawarammana,

2017). KFD doesn't have any specific treatment policy it is limited only to treat the symptoms (Jang *et al.*, 2000).

### Case History

14 yr old girl presented with complaints of fever and weight loss for 5 months. The child was apparently normal few months back and her weight was 40 kg after which the parents noted evening rise of temperature which was initially managed with symptomatic and supportive measures. She also complaint of arthralgia with features of arthritis involving mainly the small joints of hands, which subsided by itself. She was having poor appetite and a significant weight loss was noted and now she weighed up to 35 kg. In view of persistent fever she was admitted in local hospital. There Mantoux test was found to be positive and was started on ATT (AKT3) which include ethambutol 800mg, rifampicin 450mg, INH (Isoniazid) 300mg was given for 10 days. There is no history of contact with TB patient. There is no history of cough or breathing difficulty. She had bilateral axillary swelling of small size with pain not associated with pus drainage. She was referred to pediatric surgery for biopsy of swelling. There was no history of altered bowel and bladder habits, loose stools, abdominal pain and also no history of oral ulcers, photo sensitivity, rash and bone pain.

At the time of admission child was a febrile with stable vitals. On clinical examination she was noted to be pale with generalized lymphadenopathy (mainly axillary LN) with mild hepatomegaly. No rashes, skin or mucosal membrane bleed or ulcers noted. Blood counts showed pancytopenia with elevated inflammatory markers (CRP-193.4 mg/ml/ESR-62mm/hr). She was started on Inj Piperacillin Tazobactam 3.5g TID after taking samples for relevant cultures. Along with piptaz she was getting paracetamol 500 mg and pantoprazole 40 mg. Her INR was found to be 1.59 so one dose of Inj vitamin K 10mg was given as stat. Serum LDH (2789 U/L) and ferritin (16686 ng/ml) were markedly elevated. LFT showed A/G reversal with increased SGOT, SGPT. RFT and urine routine and serum electrolytes were normal. Peripheral blood smear showed leucopenia, normocytic normochromic anemia and platelet at lower limit of normal. Blood cultures were also sterile. Chest X-Ray showed hilar prominence. USG Abdomen was done which showed minimal free fluid noted in the abdomen. ECHO was done which was normal with no evidence of any vegetation.

In view of clinical picture and lab values a possibility of autoimmune disorder with MAS or hematological malignancy was considered initially. Pedi-

atric surgery consultation was availed for LN biopsy. Bone marrow aspiration and biopsy was also done. Pediatric rheumatology consultation was availed and agreed to continue with above treatment. Lymph node biopsy showed paracortical hyperplasia with foamy histiocytes possible kikushi lymphadenitis in xanthomatous phase. Considering the clinical features, age and sex RA factor (<8 IU/ml), serum complements and dsDNA (12.0 IU/ml) values were also sent and was normal. ANA was positive. Marrow biopsy and IPT reports were normal. She remained a febrile during hospitalization and IV Piperacillin Tazobactam were continued for 7 days. She had a spontaneous remission of the disease. Serial monitoring of counts and inflammatory markers were done and showed a progressively improving trend. Serum ferritin and LDH levels decreased drastically. She was discharged with Cap. fefol for 1 month and pantoprazole 40 mg for 7 days.

### RESULTS AND DISCUSSION

Kikuchi Fujimoto disease is an uncommon cause of lymphadenopathy which is often rare and benign histiocytic necrotising lymphadenitis (Singh and Shermetaro, 2019; Ifeacho *et al.*, 2008). The clinical presentation of KFD is cervical adenopathy associated with fever and night sweats which often doesn't respond to antibiotics and other treatment methods. Other clinical features are also present which include chills weight loss and systemic complaints (Hamdan *et al.*, 2002). Rare symptoms include upper respiratory symptoms, nausea, anorexia, vomiting, weakness, weight loss, chills, cutaneous rash, arthromyalgias (Papaioannou *et al.*, 2002). In our case she was of 40kg and now it reduced to 35 kg approximately 5 kg was lost also with complaints of fever mostly at evening time.

The etiology and pathogenesis of Kikuchi Fujimoto disease is not known and there are no criteria developed for diagnosis of this (Lelii *et al.*, 2018). The most common finding is leukopenia, followed by atypical lymphocytes; however, in majority of patients Complete Blood Cell count is normal. Erythrocyte Sedimentation Rate tends to be elevated and also abnormalities in liver enzymes and an elevated LDH (Humphreys *et al.*, 2018). In our case erythrocyte sedimentation rate was 62mm/hr and her SGOT & SGPT levels were elevated (1882 IU/L & 319 IU/L). The level of LDH was 2789 U/L which was highly elevated.

KFD is usually diagnosed by histopathological studies of lymph nodes which is characterized by focal necrosis in cortical and paracortical areas. Histopathologic studies cannot be ruled out at any

time so patients with KFD need to be evaluated for SLE and should have a long term follow up because of possible onset of SLE (Kampitak, 2008). Three main patterns are identified in lymph node biopsy i.e proliferative, necrotizing and xanthomatous (Ifeacho et al., 2008; Nair et al., 2020). Among these three types xanthomatous type is often rare with abundant foam cells. In our case Lymph node biopsy showed paracortical hyperplasia with foamy histiocytes- possible kikuchi lymphadenitis in xanthomatous phase.

KFD is generally self limiting disease which rarely requires treatment and often resolves within months. Only supportive treatment is present which include antipyretics and analgesics such as paracetamol and NSAIDS. For severe cases and also when supportive measures fail to resolve symptoms corticosteroids are been used (Dalugama and Gawarammana, 2017). Steroid therapy is recommended only when KFD is associated with hemophagocytic syndrome, SLE, or other rheumatic disorders (Famularo et al., 2003; Nair et al., 2020). In our case patient was prescribed with Piperacillin Tazobactam for 7 days and tablet Paracetamol was also given.

## CONCLUSION

Kikuchi Fujimoto disease is often a rare disease that initially appears with fever and lymphadenopathy along with other symptoms which include respiratory infection, less frequent chills, night sweats, arthralgia, rash and weight loss. The etiology of this disease is unknown and the recurrence rate is also low. It is often misdiagnosed with tuberculosis, lymphoma, ASOD, SLE etc. The final diagnosis is made by lymph node biopsy. It is important to diagnose the disease at initial phase in order to minimize the potentially harmful and unnecessary evaluation and treatment. Supportive care is the treatment available for KFD disease. For severe condition corticosteroids is considered. Usually the symptoms resolves in 6 months.

## Conflict of Interest

The authors declare that there is no conflict of interest for this study.

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