

## Kikuchi-Fujimoto Disease (KFD): A Case Report.

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### CASE REPORT

A 12 year old boy from Bangalore was admitted in Fortis Hospital, Shishadripuram, Bangalore with multiple neck swellings, fever, and polyarthritis of 12 days duration. On clinical examination there was bilateral, medium to large sized, mobile and tender cervical lymphadenopathy. Larger node was the right posterior cervical (3x4cm). Other nodes were of medium size (2x2cm) at left posterior cervical, left posterior auricular, left and right submandibular and right supraclavicular regions. Lymph nodes were not palpable in other parts of the body.

His respiratory, cardiovascular, and neurological examination was normal. Throat examination was also normal. His CBC was normal. ESR showed only mild elevation (22mm/1<sup>st</sup> hr). Renal and LFT were normal. Blood and urine cultures were negative. Mantoux (Tuberculin) test was negative (induration 3 mm). Ultrasound abdomen & chest X-ray were normal. ANA and anti-dsDNA antibodies were also negative. FNAC (Fine Needle Aspiration Cytology) from right posterior cervical node showed features of non-specific lymphadenitis. Keeping in view of the above picture, initial diagnosis was PUO (Pyrexia of Unknown Origin) with non-specific lymphadenopathy.

The patient was given paracetamol and broad-spectrum antibiotic. In spite of one week of antibiotic therapy, there was no response and patient continued to have fever and persistent lymphadenopathy. Finally, lymph node biopsy was done and histopathology revealed the diagnosis of KFD (Kikuchi-Fujimoto Disease). Biopsy showed patchy areas of necrosis, proliferation of pale histiocytes, increased number of apoptotic cells, cellular debris and nuclear dust (karyorrhexis). Immunohistochemistry showed CD68 positive

histiocytes. CD20 positive B-cells were also present in the lymph node areas. Patient was given corticosteroid (prednisolone) orally along with paracetamol. His fever subsided in one week and the lymph nodes regressed in 6 weeks.

(Keywords: Kikuchi-Fujimoto Disease, Bangalore, India, diagnostics, lymph nodes, corticosteroid therapy)

### DISCUSSION

Kikuchi first described the disease in 1972 in Japan (1). Fujimoto and colleagues independently described the disease in the same year (2). Kikuchi-Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis is an uncommon, idiopathic, generally self-limited cause of lymphadenitis (1, 2).

KFD is extremely rare disease (3). Its incidence has been reported worldwide with a higher prevalence among Japanese and other Asiatic individuals (3). It is observed that KFD is more common in females compared to males with male to female ratio of 1:4 (3, 4). The present case is a male patient. People under 30 years of age are more affected by this disease than any other age group (3,4). The present case is a 12 year old boy. KFD in children is reported by several workers (5,6,7,8).

KFD most often presents with cervical lymphadenopathy which may be tender and can be accompanied by fever, upper respiratory tract symptoms (3). Less common symptoms include arthralgia, skin rashes (9, 10, 11), weakness and night sweats (3). Weight loss, diarrhea, anorexia, chills, nausea, vomiting, chest and abdominal pain have also been reported (3, 10). Some patients may also have hepatosplenomegaly (10, 12, 13). Some cases exhibit ophthalmic

complications like papillary conjunctivitis, panuveitis, etc. (14, 15).

The present case exhibited cervical lymphadenopathy, polyarthritis and fever. The exact etiology and pathogenesis of KFD is unknown. The clinical presentation, course, and histological changes suggest an immune response of T cells and histiocytes to an infectious agent. Numerous inciting agents have been proposed, including Epstein Barr virus (EBV) (9,16), human herpesvirus 6 and 8 (17), HIV, parvovirus B19 (18), paramyxoviruses, parainfluenza virus, *Yersinia enterocolitica*, and *Toxoplasma* (19). There are several reports suggesting an association between KFD and SLE (Systemic Lupus Erythematosus) (3). However no convincing evidence is available to confirm such association (3).

The pathogenesis of KFD is still not fully understood. It is supposed that the primary event may be the activation of T lymphocytes and histiocytes. Proliferating T cells enter the cycle of apoptosis, which may form the areas of necrosis in lymph nodes and then the cellular debris is removed by histiocytes (20). The immunophenotype of KFD is primarily composed of mature CD8-positive and CD4-positive T lymphocytes. High rate of apoptosis is also seen among lymphocytes and histiocytes. The histiocytes express histiocyte-associated antigens such as lysozyme, myeloperoxidase (MPO) and CD68 which can be detected by immunohistochemistry. Plasmacytoid monocytes are also positive for CD68 but not for myeloperoxidase (MPO) (3, 21).

No specific treatment is available for KFD. Treatment is generally supportive. Non-steroidal anti-inflammatory drugs (NSAIDs) may be used to alleviate lymph node tenderness and fever. The use of corticosteroids (prednisolone) has been recommended in severe form of disease (22). The present case also received prednisolone with good results. Intravenous Immunoglobulin has also been tried with some success (23). The disease usually runs a benign course and the condition is self-limiting, usually resolves in several weeks to months (3). The disease has a recurrence rate of 3 % - 4 % (19, 24, 25).

Clinically KFD may mimic lymphoma or SLE. Therefore, a careful histopathological examination is necessary in arriving at the diagnosis. Hence, early recognition of the disease is of importance in

minimizing potentially harmful and unnecessary evaluations and treatments (3).

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## SUGGESTED CITATION

Khan, M.Q. 2015. "Kikuchi-Fujimoto Disease (KFD): A Case Report". *Pacific Journal of Science and Technology.* 16(2):216-218.

