

KIKUCHI FUJIMOTO DISEASE IN YOUNG CHILDREN – A CASE REPORT



Dr D S Malik

Director - Gen. & Lap Surgery

Dr S D Sharma

Director & HOD - Paediatrics & Neonatology
Eternal Hospital Jaipur, India

OBJECTIVE :

- To discuss about clinical presentation and management of a rarely diagnosed kikuchi disease

INTRODUCTION :

- **Kikuchi disease**, also known as histiocytic necrotizing lymphadenitis was first described by Dr Masahiro Kikuchi in 1972 and independently by Y. Fujimoto in Japan [1]
- Kikuchi Fujimoto disease is a very rare disease, mainly seen in Japan. Isolated cases are reported in North America, Europe, Asia, England, and at least two cases in New Zealand. after inguinal hernia repair with a new dynamic, self fixating Proflor mesh

ETIOLOGY :

- Many theories exist about the cause of KFD. Microbial/viral or autoimmune causes have been suggested [2]
- Mycobacterium szulgai and Yersinia and Toxoplasma species have been implicated. More recently, growing evidence suggests a role for Epstein-Barr virus, as well as other viruses (HHV6, HHV8 [Human Herpes Virus], Parvovirus B19, HIV and HTLV-1 [Human T Lymphotropic Virus]) in the pathogenesis of KFD.

However, many independent studies have failed to identify the presence of these infectious agents in cases of Kikuchi lymphadenopathy.

CLINICAL FEATURES:

- The patient usually presents with fever, enlargement of the lymph nodes (usually posterior cervical) skin rashes, and headache
 - Lymph node size is usually 1-2 cm, but up to 7 cm has been reported in literature. Occasionally, the supraclavicular and axillary lymph nodes become swollen.
- Rarely, enlargement of liver and spleen and nervous system involvement resembling meningitis are seen.

DIAGNOSIS / DIFFERENTIAL DIAGNOSIS :

- It is diagnosed by lymph node excision biopsy
- The differential diagnosis of Kikuchi disease includes systemic lupus erythematosus(SLE), disseminated tuberculosis, Infectious Mononucleosis, lymphoma, sarcoidosis and viral lymphadenitis [3]

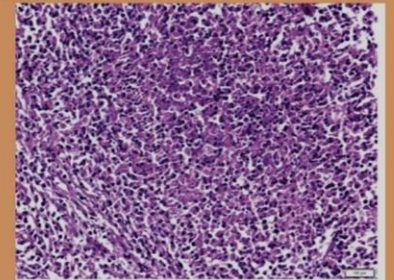
TREATMENT :

- No specific cure is known.
- Treatment is largely supportive. Nonsteroidal anti-inflammatory drugs (NSAIDs) are indicated for tender lymph nodes and fever, and corticosteroids are useful in severe extra nodal or generalized disease [4]

CASE REPORT :

- A 11 year old child was admitted with the complaints of moderate to high grade fever with chills for 1 day and swelling left submandibular region for last one month.
- O/E there was a large tender lymphnodal swelling in left submandibular region about 5x2 cm, size. Besides there were few small lymph nodes in vicinity, almost matted.

- USG neck revealed large necrotic lymph node in left submandibular region closely abutting ipsilateral submandibular gland – likely tubercular.
- All other investigations done to find out the cause of fever were negative except that Dengue ELISA NS1 was positive & the patient was managed accordingly.
- Cervical lymph node (left submandibular region) excision biopsy done.



Histopathology report show lymph nodal tissue with paracortical, well circumscribed necrotic lesions with karyorrhectic debris and fibrin deposits. There are numerous plasmacytoid monocytes, phagocytic and foamy histiocytes suggestive of Histiocytic necrotizing lymphadenitis, likely Kikuchi disease.

CONCLUSION :

All clinicians should keep this disease in mind and should be suspected when someone presents with fever, maculopapular rash, and persistent tender cervical lymphadenopathy that does not improve with antibiotics, and maculopapular rash

REFERENCES :

- 1 Masab M, Farooq H. Kikuchi Disease. Clin Rheumatol. 2017 Jun. 27(8):1073-5.
- 2 Jaseb K, Nameh Goshay Fard N, Rezaei N, Sadeghian S, Sadeghian S. COVID-19 in a case with Kikuchi-Fujimoto disease. Clin Case Rep. 2021 Mar. 9 (3):1279-1282.
- 3 Dorfman RF, Berry GJ. Kikuchi's histiocytic necrotizing lymphadenitis: an analysis of 108 cases with emphasis on differential diagnosis. Semin Diagn Pathol. 1988 Nov. 5(4):329-45.
- 4 Singh YP, Agarwal V, Krishnani N, Misra R. Entesitis-related arthritis in Kikuchi-Fujimoto disease. Mod Rheumatol. 2008 May 10. epub ahead of print.