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Case Report

Kikuchi-Fujimoto Disease: A Rare Case of Pyrexia of Unknown Origin

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Abstract

A young Male was referred to a clinician for complaint of fever of short duration with nonspecific joint pain and treated for 1 week it but after some time again presented with same complaints with severe epigastric tenderness and vomiting. After extensive work up; generalised lymphadenopathy was found with elevated ESR and C- Reactive protein with all negative Viral, Bacterial and Auto immune markers on blood & serological investigations. At last when cervical lymphnode biopsy was performed; It shows Histiocytic necrotizing lymphadenitis (Kikuchi-Fujimoto Disease).

***Key Words*:** *PUO, Cervical Lymphadenopathy, Histiocytic Necrotizing Lymphadenitis, Lymphoma, Sjogren's Disease.*

Background & Discussion

Kikuchi-Fujimoto disease, a rare form of necrotizing lymphadenitis, is self- limiting disorder of unknown Etiology. Kikuchi-Fujimoto disease or Kikuchi disease described originally in young Japanese women [3] is a rare benign cause of fever and lymphadenopathy usually involving cervical lymph nodes. The disease has been reported worldwide in both genders across ethnic and age groups.

The onset of KFD could be intense or sub-acute & Symptoms can last up to weeks and, in some cases, even months. Unilateral or bilateral cervical tender lymphadenopathy associated with low-grade fever typically occurs in the posterior cervical triangle. However, lymph node sizes can range from 5 cm to 6 cm and are rarely larger than 6 cm. The disease can cause generalized lymphadenopathy [4], though it is pretty rare.

A low-grade fever may be reported in 50% of KFD patients, along with respiratory symptoms such as cough and headache. Less frequent symptoms of KFD include skin rash, weight loss, nausea, vomiting, sore throat, and night sweats. Apart from fever and tender cervical lymphadenopathy [4], leukopenia has been detected in 50% of KFD patients. Atypical lymphocytes, similar to those identified in EBV infection, are seen in peripheral blood studies. Extra nodal involvement is relatively infrequent in KFD. Splenomegaly and hepatomegaly can also occur in few cases. The bone marrow and nervous system involvement have also been reported among a few KFD patient.

Clinically, as compared to adults, children are more prone to severe and protracted fever and have a higher risk of lymph node necrosis. The disease is often linked to an autoimmune [1,2,5] or viral Etiology, resulting in an overactive T-cell-mediated immune response. It is sometimes related to Yersinia enterocolitica and Toxoplasma Gondi infections.

Additionally, Epstein-Barr virus (EBV) and cytomegalovirus (CMV) may be considered in the pathogenesis of KFD. Other differentials of KFD include cat scratch disease and AIDS. Low-grade fever and localized lymphadenopathy of KFD might be mistaken for tuberculosis. It serves as the most prevalent preliminary diagnosis for any long-term non-tender lymphadenopathy. Both illnesses also cause nocturnal sweats and weight loss. Fine-needle aspiration cytology (FNAC) is less invasive and cost-effective and the diagnostic time is quicker but its accuracy is only 50%.

However, upon observing atypical clinical symptoms, it is recommended to perform a histological assessment of the affected lymph nodes for diagnosis. Based on the foregoing discussion, a viral or unknown infectious pathogen triggers an inflammatory response and causes KFD.

Treatment

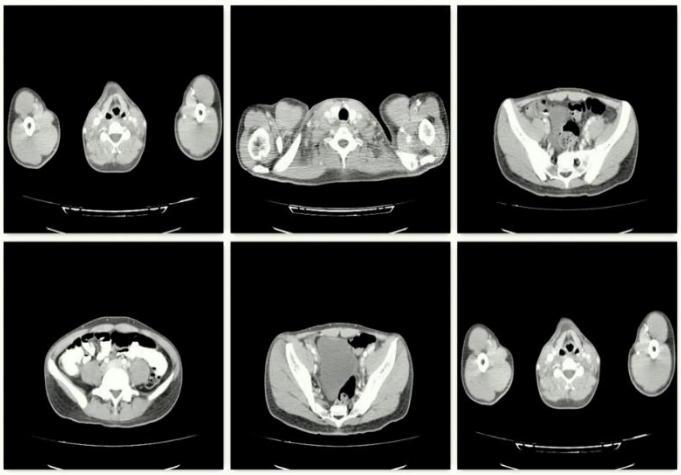
No specific treatment is available for Kikuchi-Fujimoto disease, as it typically follows a self-limited course with spontaneous resolution occurring within 1 to 4 months. Supportive management is the mainstay of treatment and involves using antipyretics and analgesics to alleviate symptoms. For patients who have a co-diagnosis of Kikuchi-Fujimoto disease and other autoimmune diseases (with SLE being the most common), appropriate treatment for the secondary autoimmune disorder is required. This may involve using agents such as corticosteroids and hydroxychloroquine.

Case Report

A 21-year boy having H/O fever, joint pain and abdominal pain for 1 week. All blood investigations with Chest X-ray, Ultrasonography of abdomen were also performed but all were normal. Blood cultures were sent; which came negative. Surprisingly fever subsided with 7 days' treatment of Antipyretics and empirical Antibiotics. But again after 3 days' fever started and this time it is with sever vomiting with epigastric pain. P/A examination was tender.

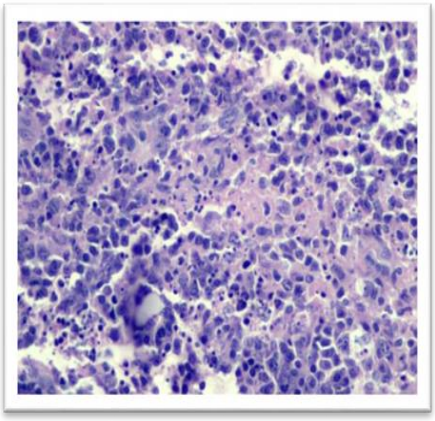
As on routine blood investigations we found CRP-167 mg/dl and ESR -77 after 1 hr. This time for thorough investigations is sent including blood Culture and widal test. CECT thorax with abdomen was done which shows multiple cervical, thoracic and abdominal lymphadenopathy. These nodes were small to medium in size. After all investigations and treatment fever and epigastric pain was not subsided; excisional biopsy of cervical lymphnode performed and it showed the following picture of Kikuchi-Fujimoto Disease.

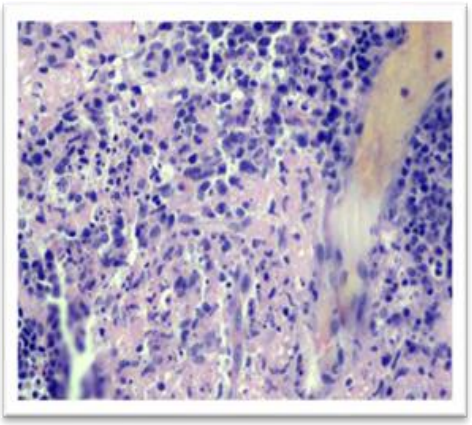
Imaging

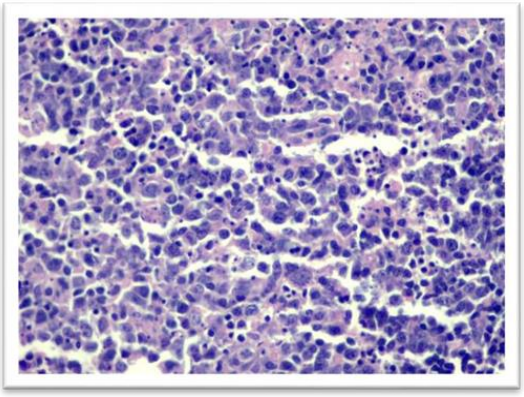


Histopathology

Figure 2: Necrotic histiocytic lymph nodes.







Conclusion

A variety of clinical presentation of KFD creates significant diagnostic and therapeutic challenges. Lymph node biopsy is best for diagnosing KFD. Investigations should be continued until the primary Etiology of the disease is found. Many clinical studies showed that KFD could occur due to an immune response to underlying autoimmune diseases, viral diseases, or bacterial infections like tuberculosis. Therefore, the diseases associated with KFD also need to be studied. Information sharing will help determine the Etiology, prognosis, pathophysiology, and definitive treatment over the long term.

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