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Kikuchi-Fujimoto Disease: A Case Report

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Abstract

Kikuchi disease, also known as Kikuchi-Fujimoto disease or Kikuchi histiocytic necrotizing lymphadenitis, is a rare, benign disorder with no known aetiology that is typically characterised by fever and cervical lymphadenopathy. It was first discovered in young women. Although the exact cause of Kikuchi disease is unknown, the histologic findings and clinical manifestations point to a T cell and histiocyte immune response to an infectious agent. Systemic lupus erythematosus and kikuchi syndrome both share similar histologic characteristics as well as sex and age propensity. The most typical clinical signs of Kikuchi illness include fever and cervical lymphadenopathy in a young woman who has previously been asymptomatic. Other symptoms include rash, arthritis, fatigue, and abdominal pain. Although no specific blood investigation is yet found to be diagnostic, the disease is confirmed by excisional biopsy of the lymph node. Here we discuss a case of a young female who presented with fever of unknown origin for a prolonged period and had been visiting multiple health centres because of no proper diagnosis.

Keywords: Kikuchi, Lymphadenitis, Biopsy

Introduction

Kikuchi disease is a rare, benign condition of unknown etiology. Although Kikuchi disease was once thought to affect young women, it has been found that the disease may present in males as well. The M:F ratio ranging from 1:4 to 1:1.6. In around 30-50% of cases, fever is the main symptom. It normally lasts a week and is of low grade, rarely lasting more than a month.[1] Although this illness has been observed in patients ranging in age from 6 to 80 years, the majority of patients are typically younger than 40 years old.[2] Most common clinical features include Lymphadenopathy (100%), Fever (35%), Rash (10%), Arthritis (7%), Fatigue (7%), Hepatosplenomegaly (3%). Other features include rigors, myalgia, arthralgia, and chest and abdominal pain.[3] Usually the disease is benign, but some cases of older patients have been reported suggesting higher risk of evolution into an autoimmune syndrome, with some patients requiring intensive care, and increased the risk of in-hospital mortality.[4] The Epstein-Barr virus (EBV), human herpesvirus 6 and 8, human immunodeficiency virus (HIV), parvovirus B19, paramyxoviruses, parainfluenza virus, Yersinia enterocolitica, and Toxoplasma have all been proposed as the inciting agents that cause the disease. Whereas, CD-8 positive T cells and histiocystes have been proposed to be involved in the pathogenesis.[5] The diagnosis is made by the histo-pathological examination of the involved lymph nodes. Early diagnosis of patients is necessary to prevent long term mistreatment on the lines of tubercular adenitis or lymphoma.

Case Report:

A 32 year old known case of hypothyroidism (on regular treatment) presented with the complaints of fever, low grade, not associated with chills or rigors and with evening rise of temperature for 1 month. It was accompanied with history of abdominal

discomfort on and off. There was no history of cough, burning micturition. There was no history of similar complaint in any of the family member. Patient's husband is a known case of pulmonary koch's treated 6 years back. For these complaints, patient had been taking medication from many health centres but was not relieved. Physical examination of all the systems was normal. Patient was subjected to multiple tests like complete blood count, chest radiograph, urine routine and microscopy, abdominal ultrasound and sputum examination which were all within normal limits. General blood picture was normal with no evidence of any parasite or toxic granules. Blood and urine cultures were obtained which were both sterile. Serology for dengue, enteric fever and malarial parasite were also negative. Stool culture was also found to be negative. Stool microscopy revealed no cyst or ova however occult blood was positive. An upper gastrointestinal endoscopy was carried out which revealed Gastric Antral vascular ectasia (GAVE). A screening anti-nuclear antibody (ANA) titre was done, which was also found to be negative. On the suspicion of Scleroderma owing to the presence of GAVE on UGIE, Anti-Scl70 antibody titre were sent which were also negative. No vegetation or effusion was found on screening echocardiography. Despite all the investigations being normal, patient was still febrile with a documented temperature of ~101F. A repeat detailed head to toe examination of the patient was done, which revealed enlarged cervical lymph node in the post auricular region on the right side. An excisional biopsy of the same lymph node was sent, which revealed features suggestive of Kikuchi Disease. The patient was counselled about the disease and was kept on acetaminophen and discharged in a stable condition. In a recent 1 month follow-up, patient had no complaints of fever or any new complaints.

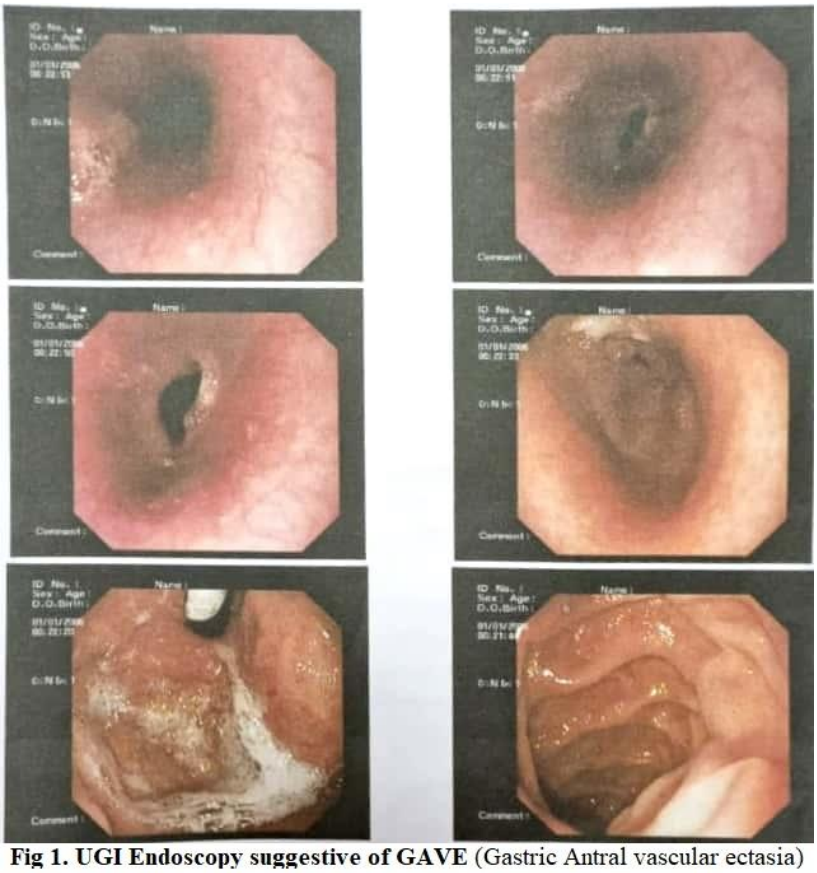

Fig 1. UGI Endoscopy suggestive of GAVE (Gastric Antral vascular ectasia)

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Received one lymph node measuring 1.3 x 1 x 0.6 cm.

Post-auricular lymph node biopsy: Features are suggestive of Kikuchis disease.

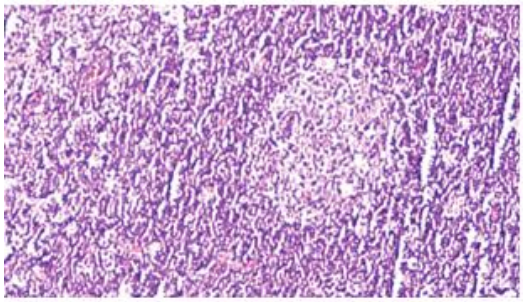

Fig 2. LN Biopsy showing reactive germinal centre with expanded paracortical area.

Discussion:

Most frequently, excisional biopsy has been advised for the diagnosis with other modalities like needle aspiration having a lower sensitivity. Microscopically, paracortical foci are frequently seen together with necrosis and a histiocytic cellular infiltration. There may be one or more foci. There may be infiltration of the capsule, and perinodal inflammation is common. The necrotizing process is frequently limited to circumscribed regions of eosinophilic fibrinoid material with an uneven distribution of nuclear debris particles. The diagnosis of Kikuchi disease does not need the presence of obvious coagulative necrosis. Around necrotic areas, there is a predominance of CD8-positive cytotoxic T lymphocytes, which may help distinguish this condition from SLE and reactive lymphoid hyperplasia.[6] Skin biopsy samples' histologic characteristics are extremely variable and non-specific.

Epidermal alteration (most frequently with necrotic keratinocytes), nonneutrophilic karyorrhectic debris, basal vacuolar change, papillary dermal edoema, and a lymphocytic infiltration are common histologic findings that may be present.[7] For Kikuchi disease, no proven effective treatment exists. Typically, symptoms and signs disappear within one to four months.[8] Patients with severe or persistent symptoms have reportedly responded favourably to treatment with glucocorticoids or high-dose glucocorticoids combined with intravenous immune globulin. Recurrent Kikuchi disease has reportedly been effectively treated with hydroxychloroquine monotherapy or in combination with glucocorticoids. In one case, a patient who was refractory to steroid therapy responded to the interleukin 1 inhibitor anakinra.[9-10] According to a report, Kikuchi disease was found associated with primary sjogren's syndrome which is an autoimmune disease.[11] It was also reported Most patients with GAVE suffer from liver cirrhosis, autoimmune disease, chronic kidney failure and bone marrow transplantation.[12] In our case, we found GAVE in a patient with Kikuchi disease. A relation between both these rare entities need to be explored.

Conclusion:

Although being a benign condition, Kikuchi disease presents as a diagnostic challenge to the physicians. The lymphadenitis may be mistreated as tubercular infection in developing country like India, where tuberculosis is quite common. Prompt and correct diagnosis is also required to prevent aggressive and wrong treatment on the lines of malignancy. Symptomatic treatment and counselling of the patients is the best way to approach management of Kikuchi Disease.

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