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CASE REPORT

FEVER WITH CERVICAL LYMPHADENOPATHY, COMMON PRESENTATION WITH RARE DIAGNOSIS-KIKUCHI FUJIMOTO DISEASE

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ABSTRACT: Fever with cervical lymphadenopathy is a very common presentation in primary care setting. Often we tend to ignore the comparatively rare diseases. If suspected early it will cut down the cost of treatment and prevent unwarranted over diagnosis of other serious conditions. Contrary to the previous case reports our patient showed protracted course of disease, which is rare in Kikuchi Fujimoto Disease (KFD).[1] **BACKGROUND:** Fever with lymphadenopathy is a rather common presentation in primary care setting. There are at least 100 differential diagnoses. More than 2/3rd patients with lymphadenopathy will have either non-specific causes or mild upper respiratory illnesses. The most common causes of benign lymphadenopathy are infectious mononucleosis, toxoplasmosis & tuberculosis, may be in the reverse order in the Indian scenario. We tend to ignore the often comparatively rare benign diseases, which are commonly missed by pathologists even. If they are suspected, the cost of treatment comes down & unwarranted over diagnosis of serious conditions is avoided.

KEYWORDS: Fever, Lymphadenopathy, Kukchi Fujimoto disease.

CASE REPORT: A 27 years old male, van driver presented with history of low grade fever for last 3 years. He then developed lymphadenopathy in cervical and sub-mandibular region. This was followed by bilateral parotid swelling. The lymph nodes were 0.5 to 1.5cm in diameter. They were all mobile and firm in consistency and non-tender. Detailed examination revealed no other abnormalities. The base line laboratory examination revealed mild leucopenia. All other parameters in CBC, LFT, RFT, RBS, HIV, ANA profile and serum ACE level were normal. CT scan of neck and chest revealed enlarged level II, III, IV lymph nodes in the neck, para tracheal and subcarinal region. Patient was provisionally diagnosed as tuberculosis and given ATT trial but he did not improve even after 2 months. FNAC & lymph nodes excision biopsy were done multiple times. Initially the diagnosis was missed & finally it showed area of necrosis with intervening preserved area. Histiocytes were present at the periphery of necrotic area. It confirmed the diagnosis of Kikuchi Fujimoto Disease. Steroids were started but he showed multiple episodes of waxing & waning of sign & symptoms. He was again admitted with increased fever & glandular swelling. After extensive discussion & review of all the available literature intravenous immunoglobulin was administered in optimal dose. 3 weeks after immunoglobulin therapy patient is showing improvement, with no fever & regression in lymph node size.

DISCUSSION: Kikuchi Fujimoto disease is a rare self-limiting disease first described in Japan in 1972. It usually presents as cervical Lymphadenopathy with or without systemic signs & symptoms. Till date most of the cases have been reported from East Asia, but recently it was reported through the world & in all races. Probably it was under-diagnosed due to its self-limiting & benign course along with less awareness. It has a slight female preponderance & affects young adults (20-30years). Mortality is rare & occurs due to systemic diseases. Recurrence is 3% only.[1]

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Pathophysiology: Cause of KFD is unknown. Most accepted being KFD result when one or more unidentified agents trigger a self-limiting auto immune response. Lymphadenitis is caused due to cytotoxic & cell induced apoptotic cell death.[2] There may be a genetic predisposition to this auto immune response. Microbial agents may be viral (Epstein-barr virus, CMV, HHV, Varicella-zoster virus, Parvovirus B19, Para influenza virus, Paramyxo virus) bacterial (Mycobacterium szalgai, yersenia, toxoplasma etc.) As research have been unable to demonstrate a clear cut & serological confirmation including demonstrating of microbial agent, it is now proposed that more than 1 pathogen may be needed to trigger the characteristic hyper immune reaction.[3]

Clinical Feature: Presenting symptoms are fever, lymphadenopathy, skin rash, and headache. Rare presentations are heapatosplenomegly aseptic meningitis, acute cerebellar ataxia, encephalitis. Lymphadenopathy is mostly cervical & involves isolated location, but other regions may be involved[4]. Fever present in most of cases along with flu like prodrome, nausea & vomiting, myalgia, arthralgia & night sweats.

Diagnosis: definitive diagnosis is only by lymph node excision biopsy. It shows area of necrosis with intervening preserved area along with scattered lymphocytes mixed with immunoblast & blood vessels. Histiocytes are present at periphery of necrotic area. CBC shows leucopenia with mild granulopenia in 20-50% patients. ESR and CRP may be elevated. LDH if increased suggests hepatic involvement. Autoimmune antibody studies have to be negative to rule out SLE.[4,5]

Treatment: Treatment includes non-steroidal anti-inflammatory drugs, corticosteroids-reverses capillary permeability and decreases polymorpho nuclear cell activity resulting in decreased inflammation. Immunosuppressant is advocated only if the disease is protracted, life threatening and rapidly progressive. M. Noursadeghi et.al used intravenous immunoglobulin in a case & reported dramatic regression of the disease. It was used as it has best side effect profile.[6]

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