KIKUCHI-FUJIMOTO DISEASE- A RARE CAUSE OF CERVICAL LYMPHADENOPATHY- A CASE REPORT FROM WESTERN INDIA

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PRESENTATION OF CASE

A female patient aged 17 years presented with complaints of fever since 25 days which was moderate to high-grade, associated with chills and mostly occurred in the evening, and lump in left lower part of the neck since 25 days. The lump was gradually increasing in size and was painful. She had no past history suggestive of SLE or tuberculosis.

On examination, patient was febrile with oral temperature of 38.8º C and pale. She had a palpable left posterior triangle node measuring 2.5 x 1 cm, which was mobile, firm in consistency and tender. No palpable lymph nodes were found in other parts of the body. Examination of cardiovascular system and respiratory system was normal. Neurological and per abdominal examination was normal.

DIFFERENTIAL DIAGNOSES

SLE, lymph node tuberculosis, lymphoma, metastatic carcinomas, sarcoidosis, infectious mononucleosis, Kimura's disease and Castleman's disease.

CLINICAL DIAGNOSIS

On the basis of history and examination, patient was suspected to have lymph node tuberculosis/ SLE/ KFD.

PATHOLOGICAL DISCUSSION

Lab investigations were as follows: Complete blood count revealed haemoglobin of 7.4 gm%; total leucocyte count of 2400/mL with 30% neutrophils, 60% lymphocytes, 5% eosinophils; platelet count was 248000/mL; ESR was 52 mm/hr. Peripheral smear showed microcytic hypochromic anaemia with dimorphism and leukopenia. Reticulocyte count was 0.7%. Liver function tests and renal function tests were normal. Blood tests for ANA, RA factor, Widal, Malaria antigen, HIV, HBsAG and anti-HCV were negative. Blood culture, urine culture and sputum culture were sterile. Mantoux test was negative. No acid fast bacilli was detected in ZN staining of sputum. Mycobacterium tuberculosis was not detected in Cartridge-based nucleic acid amplification test of sputum. Chest x-ray, abdominal ultrasound and ECG were normal.

Ultrasound of neck revealed multiple enlarged lymph nodes in bilateral levels II, III, left levels IV, V, largest measuring 2.7 x 0.9 cm in left level V which appeared necrotic. Report of fine needle aspiration cytology of cervical lymph node showed polymorphous population of lymphocytes in various stages of transformation with numerous histiocytes with few lymphohistiocytic clusters seen in the background. There was no evidence of granuloma/ atypia/ malignancy. Findings suggested reactive lymphadenitis. Biopsy of cervical lymph node showed histiocytic necrotising lymphadenitis suggestive of Kikuchi-Fujimoto disease. No acid-fast bacilli were detected on ZN staining. No malignancy. Mycobacterium tuberculosis was not detected in Cartridge based nucleic acid amplification test of lymph node biopsy specimen.

On the basis of history, examination findings and investigations, patient was diagnosed as a case of Kikuchi-Fujimoto disease.

DISCUSSION AND MANAGEMENT

Patient was managed with oral prednisolone at a dose of 1mg/kg, which was gradually tapered. Patient improved symptomatically and a repeat ultrasonography of neck performed after 1 week showed a decrease in lymph node size.

Kikuchi-Fujimoto disease also known as histiocytic necrotising lymphadenitis was described in Japan in 1972 by Dr. Kikuchi[1] and in the same year by Dr. Fujimoto and colleagues.[2] The disease has a worldwide distribution, but is most frequent in Japanese and East Asian population. The exact incidence is unknown, but it is rare in India. The aetiology of the disease is not clearly understood. It probably has infective or autoimmune aetiology. Epstein-Barr virus,[3-5] herpes simplex virus, human herpes virus types 6, 7 and 8,[5-7] HIV, HTLV[4] and parvovirus B19[4,8] are possible aetiological agents, but none has been confirmed. Infection with Yersinia enterocolitica,[9] Brucellosis,[10] Bartonella henselae[11] and Entamoeba histolytica[12] have been associated with development of KFD, but none has been established as aetiological agent. Many patients with KFD suffer from various autoimmune disorders with systemic lupus erythematosus being the most frequent.[13] There are reports of KFD patients developing SLE later in their life.[14] Certain human leukocyte antigen class II genes are more common in KFD patients. These genes are more common in Japanese population.[15]

KFD mostly affects young adults < 30 yrs. old, male-to-female ratio is almost equal, although some studies suggest female preponderance.[16] Familial cases of KFD have also been reported.[17] KFD commonly presents with multiple enlarged tender cervical lymph nodes associated with fever and flu-like symptoms. Skin is the most common extranodal organ affected. Skin manifestations include maculopapular rash, malar erythema, nodular lesions and plaques.[18]

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There is no confirmatory test for KFD. Majority of cases have a normal haemogram. Many cases have leukopenia, neutropenia and anaemia. ESR, LDH and ALT are elevated in some patients.[19-20] 25% cases of KFD have atypical lymphocytes in peripheral smear.[21] Imaging studies cannot differentiate KFD from other causes of lymphadenopathy and seldom provide any conclusive diagnostic results. Fine needle aspiration cytology of lymph node can be helpful in diagnosis, but its accuracy is only 56%.[22] Diagnosis can be established by lymph node biopsy. Histological findings include distortion of normal nodal architecture. Nodules may be seen in cortex and paracortex with coagulative necrosis and abundant karyorrhectic debris. Other histopathological features include histiocytes in peripheral parts of necrotic areas, plasma cells may be present and neutrophils are usually absent. On immunohistochemical staining histiocytes expressing CD68, myeloperoxidase and CD4 are abundant. Plasmacytoid dendritic cells expressing CD123 are also seen.[23]

KFD is a usually a self-limiting condition. Treatment is mainly supportive. NSAIDS are used to alleviate pain and fever. Corticosteroids can be given in severe cases.[24] Other treatment options include hydroxychloroquine[25] and intravenous immunoglobulins.[26] Relapse rate in KFD is about 3 - 4%.[27]

Kikuchi-Fujimoto disease, although an uncommon cause of cervical lymphadenopathy and fever should be considered in differential diagnoses, as its treatment significantly differs from other causes of cervical lymphadenopathy. Lymph node biopsy can aid in early diagnosis of KFD and avoid unnecessary investigations and treatment.

FINAL DIAGNOSIS

Kikuchi-Fujimoto Disease.

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