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Original Research Paper

Medicine

KIKUCHI-FUJIMOTO DISEASE IN A YOUNG FEMALE – A RARE CASE REPORT FROM NORTH INDIA

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ABSTRACT Kikuchi-Fujimoto disease is a rare benign disease of unknown etiology. It is also called histiocytic necrotizing lymphadenitis which predominantly affects cervical lymph nodes. It has a greater tendency to affect young females. Clinically Kikuchi-Fujimoto disease may mimic with tuberculosis, systemic lupus erythematosus (SLE) and lymphoma, therefore excision biopsy of lymph node should be used to confirm the diagnosis of Kikuchi-Fujimoto disease. We report a case of 19 years old female patient, presented with high grade fever, multiple joints pain and enlarged cervical lymph nodes. Her diagnosis was conformed as Kikuchi's disease after lymph node biopsy. She was given symptomatic treatment and she recovered thereafter.

KEYWORDS : Kikuchi-Fujimoto disease; Histiocytic necrotizing lymphadenitis; Systemic lupus erythematosus.

INTRODUCTION

Kikuchi-Fujimoto disease is a rare, benign, idiopathic and self-limiting disease which mainly involves cervical lymph nodes[1]. It mainly affects young females with male to female ratio of 1:4[2], but it can also affect elder and pregnant women[3]. It was first described by Japanese scientists Kikuchi and Fujimoto in 1972[1,4]. Although etiology is still unknown but causative agent may be virus or bacteria. Kikuchi-Fujimoto disease may be misdiagnosed as tuberculosis, lymphoma and other autoimmune diseases; therefore it is essential to recognise this condition so that redundant investigations and unnecessary treatment can be avoided.

CASE REPORT

A 19 years old female was admitted to our hospital with complaints of high grade fever (103^0 F) with rigors and chills (on and off) since 1 month along with facial flushing, multiple joint pain (bilateral shoulder, knee, elbow and ankle) and cervical lymphadenopathies. There was no joint swelling, recurrent oral ulcer, raynaud phenomenon, diurnal variation, cold, cough, burning micturation, vaginal discharge, ear discharge, diarrhoea, chest pain and dyspnoea. There was no history of weight loss, no previous history of tuberculosis or contact with tuberculosis. The patient was initially treated as tuberculosis elsewhere but she did not responded well. On examination, she had enlarged lymph nodes in right supraclavicular (3x5cm), both anterior cervical (2x2cm) and both submandibular (1x1cm) regions. Lymph nodes were discrete, firm, tender and non-matted. The examination of other systems were found to be normal.

During routine investigations, her haemoglobin was 10.8 g/dL, ESR was 16 mm in 1st hr, WBC count was 8000/ µL with normal differential count, Mantoux test was negative (3mm), sputum for A.F.B. (Acid Fast Bacilli) was negative, HIV serology was negative, liver and renal function tests were normal, urine and blood culture were negative, ANA and ds DNA were negative, chest radiography and ultrasonography of abdomen was within normal limits. Fine needle aspiration cytology (FNAC) of cervical node showed features suggestive of reactive lymphadenitis. Biopsy of cervical lymph node was done which showed focal areas of necrosis which is well-circumscribed and paracortical with abundant karyorrhectic debris along with histiocytes. These histological features supported the diagnosis of Kikuchi-Fujimoto disease. (Figure 1 and 2). Patient was treated symptomatically for few weeks and subsequently her lymph nodes were regressed. She was routinely followed up for monitoring her cervical lymphadenopathy.

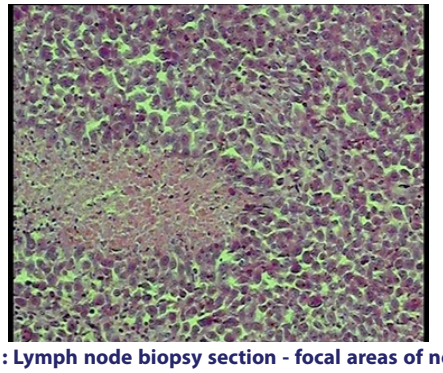

Figure 1: Lymph node biopsy section - focal areas of necrosis, collection of histiocytes with abundant apoptotic cells and karyorrhectic debris. (H&E)

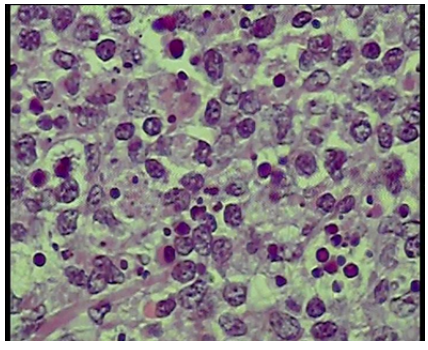

Figure 2: Lymph node biopsy section - prominence of apoptotic cells, cellular debris and nuclear dust (karyorrhexis). (H&E)

DISCUSSION

Kikuchi-Fujimoto disease is a rare and benign disease which is frequent in Asia. The most common clinical presentation of Kikuchi's disease is cervical lymphadenitis. It may occur with or without systemic manifestations[5,6]. The systemic manifestations may be fever, upper respiratory tract symptoms, arthralgia, skin rashes, weakness, weight loss, anorexia, nausea, vomiting, diarrhoea, night sweats, chest or abdominal pain and hepato-splenomegaly (rarely). Patients may also present with facial erythema, erythematous papules, plaques, nodules and ulcers[7].

There may be a nonspecific hyper-immune response to a variety of stimuli such as infectious, chemical agents and it may represent an

augmented T-cell mediated immune response in a genetically susceptible individual[8]. Viruses like Epstein-Barr virus, human herpes virus 5, human immunodeficiency virus, parvovirus B 19, paramyxo and parainfluenza viruses and also toxoplasma and other bacterial agents like Yersinia enterocolitica, Bartonella, Brucella may be the possible etiology[9]. The autoimmune mechanism may be the cause because there are reports which suggest a relationship between Kikuchi's disease and systemic lupus erythematosus (SLE)[10]. Patient may have both diseases at the same time[11]. Therefore viral lymphadenitis, systemic lupus erythematosus, tuberculosis and non- Hodgkins lymphoma may be the differential diagnosis of Kikuchi-Fujimoto disease.

CONCLUSION

The Kikuchi-Fujimoto disease is self limiting disease which usually resolves in a number of weeks to months[12]. The chance of recurrence of disease is 3% to 4%[9]. The treatment of Kikuchi's disease is symptomatic and supportive. Analgesics and anti-pyretic can be used to relieve symptoms. Corticosteroids and intravenous Immunoglobulin can also be tried for aggressive clinical course[13,14]. Therefore for the diagnosis of Kikuchi's disease a comprehensive history, complete physical examination and appropriate investigations are necessary. The early detection of the disease can be helpful in reducing superfluous evaluations and management. There should be a regular monitoring of patient for occurrence of autoimmune diseases.

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