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Kikuchi- Fujimoto Disease – A Rare Presentation

KEYWORDS: Kikuchi – Fujimoto disease, cervical lymphadenopathy, necrotizing histiocytic lymphadenitis

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ABSTRACT *Kikuchi –Fujimoto disease (KFD) is a rare benign self limiting disorder presenting predominantly in young females with cervical lymphadenopathy. Aetiology is not certain though autoimmune and infective causes have been suggested. The histological examination of lymph node is diagnostic. We report a case of a 25 yr old female who presented with fever, cervical lymphadenopathy and weight loss since six weeks. There was some relief with antibiotics and antipyretics. But the symptoms recurred with same severity after two weeks. Excision biopsy of lymph node was done which was suggestive of Kikuchi's disease. After initiation of steroids, there was dramatic response and complete recovery in 3 weeks.*

INTRODUCTION

Kikuchi's – Fujimoto disease (KFD) or necrotizing histiocytic lymphadenitis is characterized by regional cervical lymphadenopathy usually accompanied by fever. The disease was first reported by Kikuchi in Japan in 1972 and also by Fujimoto in same year[1-2]. It has a higher incidence among Japanese and other Asiatic population but a worldwide distribution is also known[3]. It usually affects individuals of age under 30 years, but it is rarely reported in children. Contrary to previous belief that it is more common in females, the various case series reports suggest that the male to female ratio is roughly equal[4].

CASE REPORT

A 25 year old female presented with complaints of multiple painful swellings in neck below and behind the left ear. The swellings were gradually progressive in size. They were accompanied by low grade fever and weight loss of 6 weeks duration. There was no history of any previous significant medical disease and she was not on any medication.

On examination, she was afebrile and haemodynamically stable. She had no pallor, icterus or rash. Multiple discrete tender lymph nodes (6 to 8 in number) were palpable involving left superficial group, deep cervical group, left posterior cervical group and left jugulodigastric lymph nodes. No other swelling was noted elsewhere on the body. Nodes were firm, freely mobile, variegated without involvement of surrounding skin; varying in size from 2.5 cm x 3 cm to 1 cm x 1 cm. No stigmata of tuberculosis were seen. Examination of throat, ear and skin was normal. Her systemic examination also did not reveal any abnormality.

Laboratory investigations revealed Hb 11.2 g/dL, TLC 2800/mm3, platelet count 1.54 lack/cumm. Significant findings were leucopoenia, raised erythrocyte sedimentation rate (ESR) - 89mm and serum lactate dehydrogenase 800 IU/L. Rest of the biochemical parameters were normal. Electrocardiogram, chest radiograph and ultrasound abdomen were normal. Mantoux test, blood, urine and stool cultures were negative. Antinuclear antibody (ANA) and anti-double-stranded deoxyribose nucleic acid (anti-dsDNA) was negative. Excisional biopsy of cervical lymph node was done which confirmed the diagnosis of Kikuchi-Fujimoto disease. (Fig. 1)

She was initially treated symptomatically with antibiotics and antipyretics. The tenderness and size of cervical nodes reduced. Fever subsided gradually over one week. After two weeks of symptomatic relief, the swellings recurred back to its original size. The patient was then treated with oral steroids (prednisolone 1mg/kg/day) in view of recurrence. Anti inflammatory drugs were continued. The patient responded dramatically and was discharged after 7 days. Steroids were tapered over next two weeks and patient was clinically stable with normal Hemogram on follow up after one month.

DISCUSSION

KFD is a benign histiocytic necrotising lymphadenitis. The exact etiology of KFD is unclear - a viral or autoimmune cause has been suggested. The role of Epstein- Barr virus, as well as other viruses (HHV6, HHV8, parvovirus B19) in the pathogenesis of KFD is controversial and yet to be proved.[3] The viral etiology is supported by its non specific, self resolving symptoms which are of slow and insidious onset. Atypical lymphocytosis, poor response to antibiotics, and also certain histopathological features like T cells favour viral etiology. KFD has been reported to occur along with SLE, and also patients of Kikuchi's disease went on to develop SLE. This supports the hypothesis that KFD may be an autoimmune disease.

The onset of KFD is acute or subacute, evolving over a period of two to three weeks. It mostly presents with cervical lymphadenopathy, consisting of tender lymph nodes that involve mainly the posterior cervical triangle. Generalized lymphadenopathy occurs rarely. Lymphadenopathy is usually accompanied by low grade fever and upper respiratory symptoms. Less common manifestations include weight loss, nausea, vomiting and night sweats. Arthralgia, skin rashes, anorexia, chest and abdominal pain have also been reported in some cases.

The diagnosis of KFD is difficult because of its non specific

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symptoms and imprecise histological findings. The laboratory investigations are also non specific and do not aid in diagnosis. Some patients have anaemia and even leucopoenia. Erythrocyte sedimentation rate, C reactive protein and serum lactate dehydrogenase are elevated. The only reliable method for diagnosis is the histological examination of lymph node excision biopsy. Fine needle aspiration is generally not sensitive, with an overall accuracy of 56.3%.[5] The presence of an enlarged lymph node with paracortical necrotic foci, which are devoid of neutrophils and surrounded by plasmacystoid monocytes, immunoblasts and cresentic histiocytes is the pathological hallmark of KFD. The immunophenotype of KFD consists of a predominance of T cells, with very few B cells. There is abundance of CD8+ T-cells over CD4+. The histiocytes express histiocyte associated antigens such as lysozyme, myeloperoxidase (MPO) and CD68. Finally, striking plasmacystoid monocytes are also positive for CD68 but not for MPO.[3]

Although KFD is rare, it must be included in the differential diagnosis of tender lymphadenopathy. It's necessary since its course and treatment differs from those of lymphoma, tuberculosis and SLE which require prompt and specific treatment whereas KFD is typically a self limiting condition and rarely requires specific treatment.

The management is mainly based on symptomatic and supportive therapy such as analgesics, antipyretics and anti-inflammatory drugs. The use of corticosteroids is not proven but has been recommended in severe extranodal or generalised KFD.[6] A high initial oral dosage of prednisolone with subsequent tapering is the proposed regimen. It is essential to keep a long term follow up of these patients, as recurrent cases of KFD can occur and also to rule out the development of SLE as there is some belief that KFD may be a precursor of SLE.[7] Recurrence rate of 3- 4% has been reported.[8]

CONCLUSION

The present case report emphasizes the need to consider the possibility of KFD while examining a young female patient who presents with fever and cervical lymphadenopathy. The constellation of clinical findings in the presence of characteristic histiocytic necrotizing lymphadenitis and pathological analysis clinches the diagnosis. Although many treatment regimens have been recommended, there is no proven therapy. Clinicians and pathologist's awareness of this disorder and early recognition leads to avoidance of unnecessary investigations, prevention of misdiagnosis and optimization of treatment results.

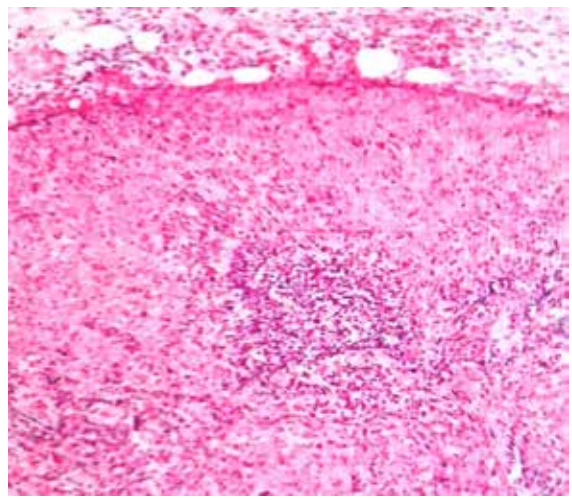

Fig 1 A section from cervical lymph node consistent with Kikuchi's disease: showing characteristic features - abundant histocytes, necrosis without neutrophils (H & E stain).

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