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A Rare Association of Kikuchi-Fujimoto's Disease With Arthritis- A Rare Case Report

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ABSTRACT

Kikuchi-Fujimoto's disease (KFD, is a subacute necrotizing lymphadenopathy of unknown etiology. It usually affects young women and has a benign course. Though it has a favorable outcome, other associated disorders like SLE, adults Still's disease and underlying malignancy has to be evaluated. A very few cases have been documented in literature regarding its association with isolated arthritis. Here we report a case of 39 year old female who presented with features of arthritis associated with cervical lympadenopathy and subsequently diagnosed as having KFD. This case report highlights the clinical features of Kikuchi-Fujimoto's disease, its associations and reinforces the importance of histo-pathological diagnosis.

KEYWORDS : arthritis, cervical lymphadenopathy, karyorrectic debris, Kikuchi-Fujimoto's disease, SLE.

INTRODUCTION

Kikuchi-Fujimoto's disease is an enigmatic, self-limited syndrome characterized by regional lymphadenopathy with tenderness, accompanied by mild fever and night sweats[1]. It was first described by Kikuchi and Fujimoto in 1972[2]. KFD has a world-wide distribution and its pathogenesis remains controversial. Some kind of viral and post-viral etiology has been suspected[3]. Several isolated cases and its association with other autoimmune disorders have been studied[4]. So far, only two cases have been reported regarding its association with arthritis as a predominant presenting symptom[5]. We present a case of KFD in with association arthritis which was managed successfully with conservative line of treatment.

CASE REPORT

A 39 year old female presented with history of fever with arthralgia since two weeks. She had pain and swelling over her left wrist and meta-carpophalangeal joints. Also she noticed mild erythematous rashes on trunk and abdomen during first two days of illness which gradually subsided. On clinical examination, posterior group of cervical lymphnodes were tender and enlarged bilaterally. There was no hepato-spleenomegaly or any other bleeding manifestations.

Laboratory investigations revealed mild leucopenia [3400/cumm], ESR of 30mm/1st hour. Peripheral smear examination, urine routine, liver and renal function tests were within normal limits. An initial diagnosis of viral arthritis was made and patient was put on non-steroidal anti-inflammatory drugs [NSAIDS]. Fever settled over several days but cervical lympadenopathy and arthralgia persisted. A differential diagnosis of connective tissue disorder was offered but all connective tissue and vasculitis screen were negative. Her Gallium scan was normal. Fine needle aspiration cytology of cervical lymphnodes was adviced. It yielded scant material comprising of few histiocytes and atypical lymphocytes which was non-contributory. Subsequently lymphnode excision biopsy was done and sent for histo-pathological opinion. On gross examination, lymphnodes were enlarged [2-5cms], cut-surface showing patchy area of necrosis [fig 1]. Microscopy showed partial effacement of nodal architecture with reactive germinal centre formation. Areas of coagulative necrosis, abundant karryorrectic debris surrounded by polymorphous population of lymphoid cells, predominantly comprising of small and large lymphocytes, histiocytes, immunoblasts and plasmacytoid cells were evident [fig 2 & 3]. No granulomas were seen. ZN stain for acid fast bacilli was negative. Underlying malignancy was ruled out. Based on these findings histological diagnosis of Kikuchi-Fujimoto's necrotising lymphadenitis was given. Patient was managed conservatively and advised regular follow up.

Discussion -

Kikuchi-Fujimoto's necrotizing lymphadenitis is typically a self-limited disease with recurrence rate of 3-4% 6. The onset is acute or subacute ranging from 2-3 weeks6. Cervical lymphodes are commonly involved [56-98%] and normally associated with pain [90%]. Generalized lymphadenopathy has been reported in 1-22% of the cases7. Our patient had bilateral tender posterior cervical lymphadenitis. Less frequently associated symptoms include weight loss, nausea, vomiting, night sweats, autoimmune disorders like SLE, polymyositis, adult's Stills disease, uveitis and pulmonary hemorrhage 8,9. Rare unusual associations like thyroiditis, large B-cell lymphoma, parotid gland involvement and haemophagocytic syndrome have also been reported10. Till date only two cases of isolated arthritis associated with KFD has been reported in the literature5, without any systemic involvement. Similar observation was made in our case, where patient presented with severe pain and swelling in the left wrist and metacarpophalangeal joints. However, arthritis has been associated with concurrent KFD and SLE, but the ANA profile was normal in our case.

There are no specific laboratory tests available for diagnosing this rare disorder as most of them will have normal picture. Leucopenia and atypical lymphocytosis has been observed in 25-50% of the cases[11]. The usefulness of fine needle aspiration cytology [FNAC], is limited and non-specific. Therefore excision lymphnode biopsy becomes mandatory for the confirmative diagnosis[12,13], especially when the clinical presentation is unusual as in our case.

The characteristic histological findings of KFD include paracortical or patchy areas of necrosis, karyorrectic debris, varied population of histiocytes, plasma cells, immunoblasts, macrophages, small and large lymphocytes and interdigitating dendritic cells. Reactive lymphoid follicles and follicular hyperplasia has been found in 10-50% of the cases. Our case had histological features consistent with that of typical KFD. Recognition of KFD is crucial as it may be mistaken for malignant lymphoma[14]. However, benign histiocytes can be differentiated from atypical lymphocytes by the presence of delicate nuclear membrane.

Currently, there is no specific treatment available for managing this disorder and most of them are managed conservatively. There are reports of excellent responses to immunoglobulins, minocycline, hydroxyquinine and low dose corticosteroid therapy[13,15]. Patients re-

quire a systematic approach with regular follow up for several years to recognize the recurrences and ongoing development of SLE or any other malignant change. Our patient was put on low dose corticosteroid therapy which was gradually tapered over several weeks. She continued with NSAIDS and advised to take adequate rest during clinical flare ups. Gradually her cervical lymphadenitis and rheumatological symptoms got resolved over 3 months. Regular follow up for once in 6 months was advised.

Conclusion –

Kikuchi-Fujimoto's disease is a benign, self-limited condition and can present with unusual features like arthritis. Therefore exact diagnosis and appropriate management requires a systematic approach for not only to rule out the underlying autoimmune diseases and malignancy but also to avoid unnecessary surgical intervention and toxic therapy.

Figures:

Fig 1- Gross photograph showing lymphnode enlargement with patchy area of necrosis

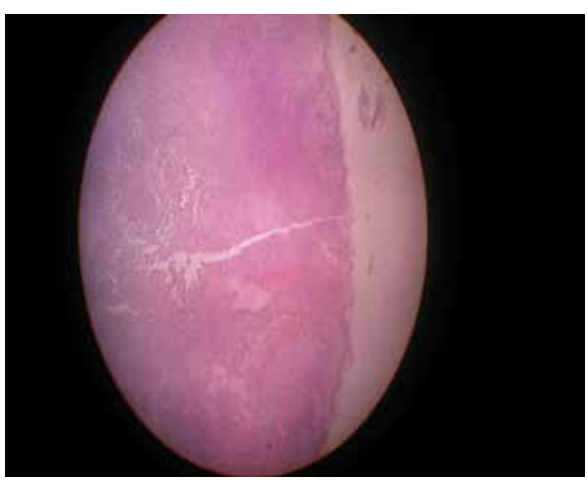

Fig 2- photomicrography of lymphnode showing partial effacement of nodal architecture and patchy area of necrosis [H & E, 100X]

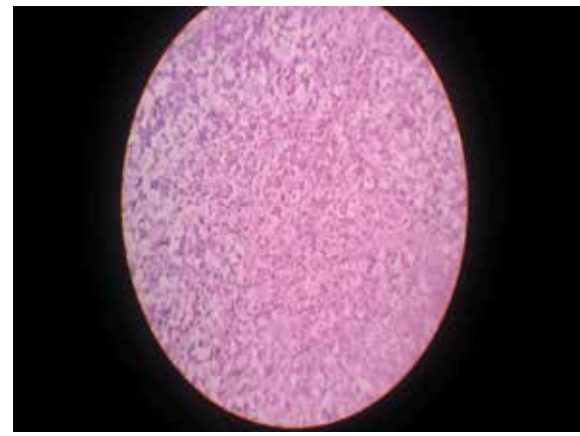

FIG 3- Photomicrography showing karyorrectic debris, varied population of histiocytes, macrophages, small and large lymphocytes.[H& E- 400X].

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