Arthritis as a Presenting Feature of Kikuchi-Fujimoto Disease: Time to Think Out of the Box in Patients with Arthritis

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Abstract

Kikuchi-Fujimoto disease (KFD) is a rare benign disorder which presents with inflammatory enlargement of the cervical lymph nodes of unknown aetiology. It is more common in young females. Its clinical presentation is quite similar to other infective, malignant and immunological disorders. We report the case of a 25-year-old female with symmetrical polyarthritis along with lymphadenopathy. Arthritis as a presenting feature is quite unusual in KFD which made this case quite challenging and difficult to diagnose.

Keywords: Histiocytic necrotising lymphadenitis, Kikuchi-Fujimoto disease, systemic lupus erythematosus

Case Report

A 25-year-old female with no other comorbid conditions presented with high-grade fever, joint pain and cervical and axillary lymphadenopathy. Clinical examination showed stable vitals with a blood pressure of 110/70 mmHg and a pulse of 88 beats/min. Enlarged posterior cervical lymph nodes and axillary lymph nodes, largest one measuring about 2x3 cm in size. Lymph nodes were mobile and mildly tender. Her metacarpophalangeal, metatarsophalangeal and proximal interphalangeal joints were painful and inflamed indicating small joint polyarthritis. There was no rash and hepatosplenomegaly.

Investigations showed haemoglobin of 9.8 g/dl with a normocytic normochromic picture and total leukocyte count of 6700/cm and an erythrocyte sedimentation rate (ESR) of 100 mm/1st h. The rest of the metabolic profile including liver and renal function and serum electrolytes were normal. Blood and urine cultures were negative. Tuberculin skin test showed an induration of 3 mm at 48 h. The ultrasound abdomen was normal. The chest radiograph was normal. Autoimmune profile including Antinuclear antibodies (ANA), Anti smooth muscle antibody (ASMA) and Anti mitochondrial antibody (AMA) was negative.

She was started on empirical antibiotics but was not responding. Cervical lymph node biopsy showed reactive lymph node with necrotising histiocytic lymphadenitis histological features of KFD [Figure 1]. No granuloma or malignant changes were seen.

Immunohistochemical staining of the histopathological specimen showed positivity for CD-20 highlighting reactive follicles and negative necrotising foci.

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How to cite this article: Nizamuddin, Ubaid A, Waheed F. Arthritis as a presenting feature of Kikuchi-Fujimoto disease: Time to think out of the box in patients with arthritis. Hamdan Med J 2020;13:55-6.
The patient was started on corticosteroids and non-steroidal anti-inflammatory drugs. A follow-up visit was scheduled at 8 weeks which showed marked improvement in the symptoms, and size of lymph nodes and joint pain had also improved a lot along with marked improvement in the laboratory values, especially ESR and quantitative C-reactive protein.

**DISCUSSION**

KFD is benign necrotising lymphadenitis presented by Dr. Masahiro Kikuchi in 1972 as a disease-causing inflammation of the lymph nodes with localised proliferation of reticular cells along with numerous histiocytes and extensive nuclear debris.[1] In the same month, Dr. Fujimoto also described a similar case in a Japanese journal.[2] Its evolution occurs over weeks to months, mainly described in Asia. It is associated with autoimmune disease, particularly with lupus erythematosus. SLE association is seen more among Asian than Caucasian population.[3]

It is most commonly associated with SLE, and one theory proposed that KFD may be a self-limiting form of SLE. The theory is supported by the fact that the histopathological findings of Kikuchi disease can be very similar to lupus lymphadenitis.[4]

KFD presents commonly with high-grade fever, enlarged lymph nodes, weight loss and sometimes with dysphagia. Dysphagia and lymphadenopathy presentation is uncommon. Knowledge about this disorder will help the clinicians and pathologists in the prompt diagnosis and appropriate treatment.[5] Cervical lymphadenopathy, especially in the posterior cervical triangle with bulky and painful lymph nodes, is commonly seen; rare cases of generalised lymphadenopathy can also be seen.[6] Clinical presentation is also accompanied by weight loss and arthralgia.[7]

Baseline investigations may show anaemia, high ESR, high C-reactive protein and lactate dehydrogenase. It is mainly diagnosed on the basis of biopsy and histopathological study of an affected lymph node. Histology of lymph node shows paracortical expansion with necrotic foci laden with karyorrhectic debris. Later stages show histiocyte replacing necrotic debris.[8] The disease is usually self-limited and does not require any specific treatment. The resolution of symptoms typically occurs within 4 months. Some patients experience recurrent episodes. Treatment with non-steroidal anti-inflammatory drugs is used to control symptoms.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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**Figure 1:** Cervical lymph node biopsy showing reactive lymph node changes with necrotising histiocytic lymphadenitis consistent with Kikuchi-Fujimoto disease