Kikuchi-Fujimoto Disease Associated with Aseptic Meningitis: A Case Report

Aseptik Menenjitle Birlikte Görülen Bir Kikuchi-Fujimoto Hastalığı Vakası

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Abstract
Kikuchi-Fujimoto Disease is usually a rare, self limiting, benign disease causing fever, neutropenia and cervical lymphadenopathy. It is especially seen in young Asian women, rarely in children. A case of Kikuchi-Fujimoto Disease in a female Bangladeshian child with aseptic meningitis is presented from Turkey. She was admitted with fever but had no pathological lymphadenopathies. Her complicated aseptic meningitis was treated successfully with short term steroid therapy.
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Key words: Kikuchi-Fujimoto Disease, meningitis, lympadenopathy, fever

Özet
Kikuchi-Fujimoto hastalığı ateş, nötropeni ve servikal lenfadenopatiyle seyreden ve özellikle genç Asyalı kadınlarda görülen bir hastalıktır. Çocuklukta nadir olarak görülür. Aseptik menenjit ile başvuran Bengaldeş'li bir kız hastada saptanması nedeniyle sunulmaktadır. Hasta inatçı ateşle müracaat etmesine karşın, lenfadenopatisi yoktur. Oluşan komplikasyonlu aseptic menenjit tablosu kısa süreli steroid ile tedavi edilmiştir. *(J Pediatr Inf 2011; 5: 77-9)*
Anahtar kelimeler: Kikuchi-Fujimoto, lenfadenopati, menenjit, ateş

Introduction

Kikuchi-Fujimoto Disease is usually a rare, self limiting, benign disease, causing fever neutropenia and cervical lymphadenopathy, seen especially in young Asian women but rarely in children (1). If it is not considered in patients with fever of unknown origin with cervical lymphadenopathy, this may result in unnecessary invasive tests. Although self limiting, rare fatal cases were reported (2).

Case

A 11 year old Bangladeshian female child who lived in Ankara/Turkey because of her father's occupation presented with persistent and intermittent fever up to 40°C of twelve days duration. She had fever and sore throat since the beginning of disease. The patient was given benzatine penicillin (1 200 000 IU) once intramuscularly and ceftriaxone for the following two days in a local hospital. Foreign travel included a brief return to Bangladesh, a trip to Canada 6 months previously and a visitto a small cattle farm near Ankara 2 weeks previously.

She appeared well; but her temperature was 39°C. There were only two non-tender lymphadenopathies 1x1, 1x0.5 cm size in the right posterior cervical region. She had no hepatosplenomegaly. The rest of the physical examination was normal.

Laboratory revealed low white cell count of 3700mm^3 with neutrophils 48%, low platelet count 141 000/mm^3, and high lactic dehydrogenease (LDH) 862 IU/L. C Reactive protein was 9 mg/dl and sedimentation rate was 36 mm/hr. Biochemical profile, thyroid function tests, quan-

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tiferon tuberculosis test with Elisa, chest x-ray and urine were normal .Initial management consisted of regular paracetemol, while awaiting results of further tests These included blood culture, viral screen, salmonella brucella agglutinin tests and autoimmune screen.

Haemotological malignancy, haemophagocytic syndrome, malaria and leishmaniasis were ruled out from the blood smear and bone marrow examination. Because of the history of a visit to a cattle farm, babeosis and lyme serology were also studied. All these culture and serological results were negative.

As she had headache and vomiting observed at the end of the first day of hospitalization, magnetic resonance (MR) was carried out and showed no abnormality except a deep posterior cervical adenopathy 3x1.5 cm in size. Cerebrospinal Fluid (CSF) analyses revealed WBC 30/mm^3, lymphocyte, glucose 42 mg/dl, protein 89.7 mg/dl. No bacteria were seen in the CSF smear and the cultures were negative. Electroencephalogram showed a diffuse slow wave abnormality. At the end of second day steroid 1 mg/kg dose IV then 1.5 mg/kg /day and acyclovir 10 mg/kg every 8 hours were started because of the deteroration in her clinical condition. She was afebrile and conscious during the first day of therapy.

Two days later, the existing lymphadenopathy from the right posterior cervical region was surgically removed. Pathology revealed a specific diagnosis of KF disease (Figure 1, 2).

Because CSF PCR for EBV and HSV were negative, acyclovir therapy was stopped. Steroid therapy tapered gradually within 2 weeks. After discharge she remained well. During follow up CBC, AST ALT and LDH returned normal to level.

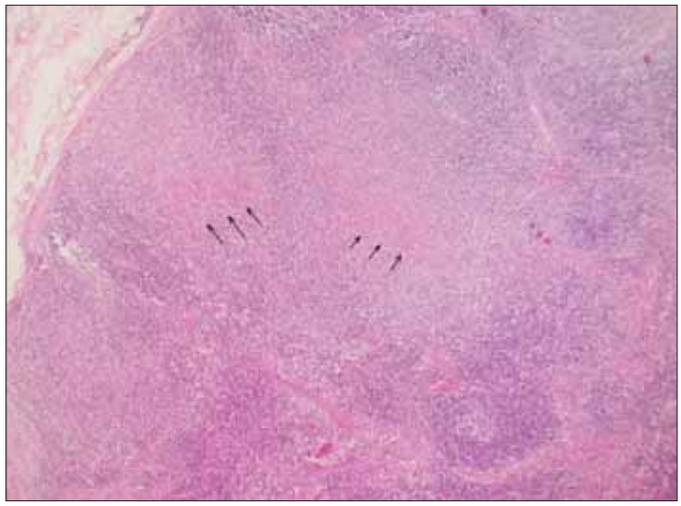

Figure 1. Necrotic areas in paracortical region region (H&E, x40)

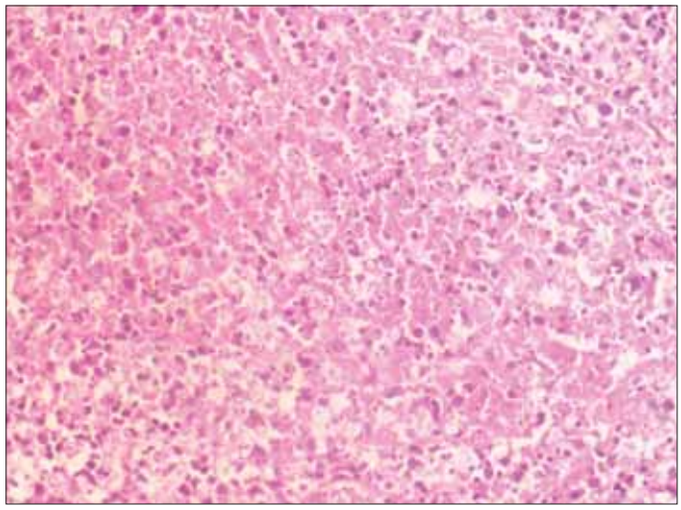

Figure 2. Karyorrhectic debris in necrotic (H&E, x400)

Discussion

KFD a self limited benign disease, identified first time in 1972 in Japan, is especially seen in eastern Asian countries in young women but rare in children.

The cause of the disease is unknown however, several infectious agents have been implicated, and it is thought that immune cells that give react as a hyper immune response (3).

Lymphadenopathy with isolated location is seen in 83% of the cases, however posterior cervical location occurs in about 65-70% of the cases (4). In our case, isolated lymphadenopathy was detected in the right posterior cervical region with MR, which was ordered for the evaluation of aseptic meningitis.

Neurological involvement is rare, but cerebellar ataxia, aseptic meningitis and encephalitis were reported as isolated cases. Nakamura et al. reported only two aseptic meningitis cases out of 69 patients (5). The mechanism of aseptic meningitis with KFD remains unclear. In aseptic meningitis cases headache was the prominent finding, but Kerning and Brudgenski signs were not reported. In our case, only severe headache and blurring of conciousness was present without positive Kernig and Brudgenski signs (6).

Our case is from Bangladesh where the disease is common. This situation was kept in mind during evaluation of persistent or unknown origin of fever. However no Kikuchi Fujimoto disease presented in two reports related to persistent fever from Turkey (7,8).

In the Kikuchi syndrome, corticosteroid therapy was only proposed in severe extranodal or generalized cases, i.e. ANA positive lupus like syndrome, hepatic involvement. Further, Jang et al. prescribed steroids in less severe cases, if the fever persists more than two weeks (9).

A 14 year old Turkish patient with lymphadenopathy without fever and a child mimicking lymphoma were reported previously (10,11).

The importance of our case is that it is the first report from Turkey in an Asian child, with no prominent pathological lymphadenopathy at physical exam, and who showed aseptic meningitis with good response to short term low dose steroid therapy.

Our case pointed out the benefit of the communication technology to the clinicians. When we enter our preliminary data to a search site as “female, Bangladesh, fever, leucopenia” the key word KFD appeared.

Finally it is well known that every geographical region has specific diseases. However, in a globalizing world, this hypothesis seems to diminish, as shown in our case. So it is worth to obtain the travelling history of the foreign patients.

Consent

Written informed consent was obtained from the patient’s father for publication of this case report and any accompanying images.

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