Kikuchi-Fujimoto Disease: A Case Report

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Kikuchi–Fujimoto disease is a rare benign condition of necrotising lymphadenitis. It usually occurs in women in early 20s to late 30s. We report a case of Kikuchi's disease that occurred in 22 years old women. She presented with fever and palpable tender cervical lymph nodes. Kikuchi-Fujimoto disease was diagnosed after lymph node biopsy. Patient was given symptomatic treatment and was cured after two weeks.

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Key words: Cervical lymphadenopathy, Kikuchi Fujimoto

Introduction

Kikuchi-Fujimoto disease (KFD) is a rare, benign and self-limiting disease involving lymph nodes. It occurs mainly in young female. This disease was first described by both Kikuchi and Fujimoto in Japan in early 1970s.[1]

It usually occurs in women in their late and early 30s and manifest usually as a posterior cervical lymphadenopathy. It resolves spontaneously over a period of several weeks to 6 months. Its initial clinical appearance is commonly similar to that of lymphoma and it can be pathologically misdiagnosed as such. Kikuchi disease might be associated with systemic lupus erythematous (SLE).[2]

Case Report

A female housewife of 22 years old from Ponchagarh district of Bangladesh presented with multiple swelling in the left side of the neck for 13 days and fever for same duration. Fever was initially low graded and intermittent, later became continuous nature. On examination, she had multiple enlarged lymph nodes in posterior cervical chain in left side of the neck. Lymph nodes were 1.5-3 cm in size, discrete, firm and tender.

Her hematological examination reports were normal, Manteaux test was negative, chest X-ray normal. FNAC from the neck swelling was done and it was non-conclusive.

Excisional biopsy of a cervical lymph node confirmed a diagnosis of Kikuchi-Fujimoto disease. Histological analysis showed collections of histiocytes with nuclear debris in the background of necrosis (Figure 1). Only symptomatic treatment was given and the patient was discharged home after two weeks in patient stay.

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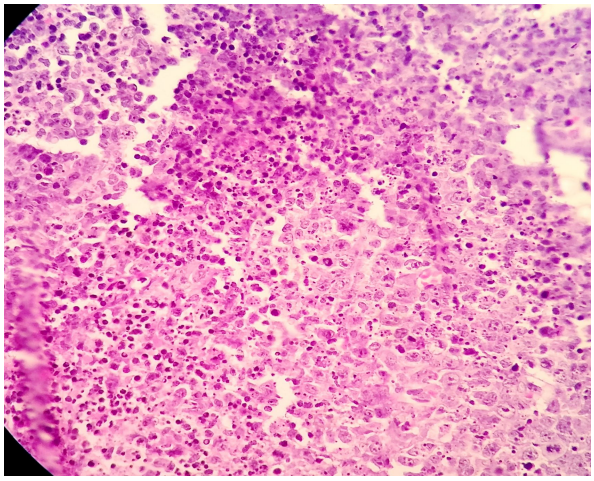

Figure 1. Histological picture of KFD:

At follow up she reports no symptoms, remains well and there are no abnormalities on clinical examination.

Discussion

Kikuchi-Fujimoto disease (KFD) was first described in 1972 in Japan. It also known as Kikuchi disease, Histiocytic necrotizing lymphadenitis, Kikuchi necrotizing lymphadenitis, Phagocytic necrotizing lymphadenitis, Subacute necrotizing lymphadenitis and Necrotizing lymphadenitis.[3]

Its etiology has not yet been fully determined; however it is believed to be a viral origin, EBV. HHV- 6 and 8 have been suggested. Raw fish was postulated as a cause, but the recent literature does not support this.[4]

An autoimmune etiology is also likely as it has been reported in association with SLE. It tends to affect a young population under 30 years of age, including children, although the later are less commonly affected. There are reported cases in older age group and pregnant women too.[5]

A definite diagnosis is made by tissue biopsy, indeed whole lymph node biopsy. Histological assessment of affected lymph nodes reveals characteristic findings. There are three main patterns identified, proliferative,necrotizing and xanthomatous .The proliferative picture is seen approximately a third of cases and has a dominant inflammatory infiltrate. Half of the cases shows necrotizing pattern and xanthomatous is rare and has abundant foam cells. Immunoblast cell changes seen in lymph nodes mimic those of malignancy and are a source of diagnostic confusion. Cellular protein structures have been noted in cytoplasm of lymphocytes and histiocytes that have also been found in those cells of patients with SLE. This adds strength to hypothesis that KFD is a self limiting SLE like disorder.[6]

The signs and symptoms of Kikuchi disease are fever, lymphadenopathy, skin rashes and headache. Rarely hepatospleenomagaly and nervous system involvement resembling meningitis is seen. Differential diagnosis includes SLE, disseminated tuberculosis, lymphoma, sarcoidosis and viral lymphadenitis.[7]

In 80% of patients, KFD is self-limited and resolves within 1 to 6 months without specific treatment. Patients with extensive systemic manifestations, pyrexia, lymphadenopathic pain or a combination of these symptoms have been treated with steroidal or nonsteroidal anti-inflammatory medications, and patients with advanced KFD may benefit with systemic prednisolone.[8]

Recurrence has been reported in some studies. Recurrence rate of KFD was 3.3% over 2 yrs in one study.[6]

Conclusion

KFD is a self-limited and typically benign lymphadinitis of unknown etiology. Confirm

diagnosis is made by lymph node biopsy. Clinical suspicions of otolaryngologic surgeon, in conjunction with pathological findings, are critical for an accurate diagnosis.

Learning Points

- Diagnosis of KFD is an important to make as treatment is conservative and this differs to the management of other conditions it may be confused with.
- KFD requires a tissue diagnosis and so lymph node biopsy should be considered in patient of cervical lymphadenopathy where KFD is suspected.
- Though rare disease, should be considered in all cases where clinical feature and FNAC cannot give any clue of common disease.

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