CASE REPORT

Cytological diagnosis of Kikuchi- Fujimoto's disease: a case report with review of literature

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ABSTRACT

Kikuchi- Fujimoto disease (KFD) is an uncommon, benign self limiting condition, which presents as cervical lymphadenopathy in young women. Fine needle aspiration cytology can help in differentiating it from other disease like tuberculosis, SLE lymphadenitis and non Hodgkin's lymphoma. We present a case of KFD diagnosed on FNAC in a twenty five year old woman.

1. Introduction

Kikuchi- Fujimoto disease (KFD) is a subacute necrotizing lymphadenopathy affecting mainly young women.[1] It is an uncommon, benign, self limiting disease presenting with firm, tender unilateral cervical lymphadenopathy. The present distribution is global, Japan and other Asian countries topping the list. Studies highlighting the role of cytology of KFD are few in literature. FNAC when performed in good hands, with a keen eye on clinical examination, may thus obviate the need for excision. We present a case of KFD diagnosed on FNAC in a young female.

Case report

A 25 year old female presented with a history of swelling in the left posterior triangle of neck since 2 months. She gave a past history of fever which was diagnosed and treated outside 2 months back. The swelling persisted since 2 months, and was not associated with cough, pain or weight loss.

On examination, a non tender, discrete left cervical swelling in posterior triangle measuring 3x3 cm, left submandibular lymph node measuring 2x1 cm and right submandibular lymph node measuring 1x1 cm was noted. Inguinal and axillary lymph nodes were normal. Examination of other systems was normal. Laboratory investigations were normal and peripheral blood smear revealed a normocytic normochromic blood picture. ESR was 50 mm/hr. Ultrasound of neck showed multiple enlarged lymph nodes at Level IIB, II, III and IV on left side, and Level IIB on right side, largest measuring 2x0.8 cm at level IIB.

Aspirate from left cervical swelling yielded 0.3 ml of pus like material. Cytology showed a polymorphous population of lymphoid cells along with histiocytes seen in singles and clusters and binucleate forms. Crescentic histiocytes were also seen. Background showed abundant necrotic and karyorrhectic debris. [Fig: 1] ZN stain was negative for AFB bacilli. Cytological features were suggestive of necrotizing lymphadenitis- kikuchi's lymphadenitis. Histopathological examination of excised lymph node confirmed Kikuchi's disease.[Fig: 2]

Discussion

KFD is a benign, rarely fatal disease presenting as unilateral cervical lymphadenopathy. The disease was first reported independently and simultaneously by Kikuchi [2] and Fujimoto et al [3] in the year 1972 from Japan. The condition was earlier commonly seen in Japan and other Asian countries, at present however the distribution is global. It is commonly seen in young women, though children may also be affected.[4] Clinically KFD may present as tender cervical lymphadenopathy, accompanied by fever and upper respiratory tract symptoms. Less common symptoms include arthralgia, skin rashes, weakness and night sweats.

Etiology of KFD is still uncertain. Infectious agents thought to play a role are EB virus, Human Herpes virus, own a good
response Cytomegalovirus and parainfluenza virus. Few authors have also reported an association between KFD and SLE. However, no convincing evidence is available to confirm such cases. [5,6] Additionally, SLE lymphadenitis should also be considered as a differential diagnosis for KFD.

Cytology reveals abundant crescentic histiocytes, plasmacytoid monocytes, variable lymphocytes, along with abundant karyorrhectic debris. Neutrophils are characteristically absent. Plasma cells are scarce or absent. [7] Crescentichistiocytes is a characteristic feature in KFD. They have eccentric placed crescentic or distorted nuclei with ingested nuclear debris. In contrast, tingible body macrophages have centrally located round to ovoid nuclei with ingested debris. [8] Karyorrhexis is also one of the exclusive features of KFD and the process may extend into the perinodal areas as well. Karyorrhecticdebris are extracellular, irregularly shaped nuclear fragments without accompanying cytoplasm. [8] They should not be confused with neutrophils which show well formed segmented nuclei with distinct cytoplasm. The cell types identified in KFD may vary with the stage of disease. Crescentichistiocytes, plasmacytoid monocytes and atypical lymphocytes along with karyorrhectic debris are seen in proliferative stage. Eosinophilic necrosis, karyorrhecticdebris, variable lymphoid cells, abundant histiocytes and plasmacytoid monocytes are seen necrotizing stage. Minimal or no necrosis along with karyorrhectic debris and foamy histiocytes are seen in xanthomatous stage. [9] Necrotizing stage accounts for most of the cases diagnosed.

Histopathology demonstrates foci of apoptotic changes in entire lymph node, mostly in the cortex and paracortical region, with abundant karyorrhectic debris and proliferation of histiocytes, plasmacytoid dendritic cells and lymphocytes in the absence of neutrophils. [10] Immunohistochemistry shows the lymphoid population to be CD3+, CD20−, CD4+, CD8+. Kikuchihistiocytes in the affected foci are CD68+ and MPO+. Studies have shown that the proliferation of CD8+ cells in the lesions initiate a killer or chemotactic mechanism which is responsible for the apoptotic sand in the lesions. [11] Transmission electron microscopy shows thickening of nuclear membrane, apoptotic bodies, and finger like network of apoptotic process in most of the cases diagnosed.

Laboratory evaluation of KFD reveals non specific findings including lymphopenia, neutropenia with atypical lymphocytes, anaemia, raised ESR, elevated levels of serum lactate dehydrogenase (LDH) and transaminases. Radiological investigation including CT and MRI may show non specific findings. Hence the need for FNAC and excision biopsy to confirm diagnosis.

Differential diagnosis to be considered are tuberculosis, SLE lymphadenitis and non hodgkin's lymphoma (NHL). Tuberculosis will show neutrophils, multinucleated giant cells and epithelioidhistiocytes. Special stains for acid fast bacilli will help to differentiate tuberculosis from KFD. SLE lymphadenitis will show presence of plasma cells and hematoxylin bodies which are absent in KFD. Serological tests will also help in diagnosing SLE lymphadenitis. Features that distinguish KFD from NHL include incomplete architecture effacement, with patent sinuses, presence of numerous reactive histiocytes, relatively low mitotic rates and absence of RS cells.

Though KFD has been known to subside spontaneously, recurrence in KFD has been reported. [1] Also KFD is thought to be a precursor for SLE, as both disease have had concurrent and coexisting disease patterns in the same patients. [1] Hence follow up may be necessary. Treatment of the disease varies from antibiotics, NSAIDs, and corticosteroids which have sh
Conclusions

KFD is an uncommon cause for cervical lymphadenopathy. However, it should be differentiated from more common causes like Tuberculosis, lymphoma and SLE also. Treatment is usually conservative. Recurrence has been reported, hence long term follow up maybe necessary.

References


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