An Unusual Presentation of Kikuchi-Fujimoto Disease on Cytomorphology: A rare case report

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ABSTRACT

Background and Objective: Kikuchi Fujimoto Disease[KFD] is an uncommon condition presenting with necrotizing lymphadenitis associated constitutional symptoms and associated autoimmune disease. Cytology is an evolving diagnostic tool in low sensitivity on this disease having many differentials often warranting biopsy. Herewith we present a rare case report of KFD with non classical mode of presentation and emphasizing the diagnostic utility of cytology in effective pick up of the diagnosis.

Case Report: A 22 year old female presented with cervical lymphadenopathy with constitutional symptoms. Unusually no associated autoimmune conditions noted with negative serology. Cytomorphology clinched the features of histiocytic lymphadenitis along with karyohexis with non necrotizing background. Subsequent biopsy proved the cytology diagnosis of KFD.

Conclusion: KFD can appear as atypical presentation without any association of autoimmune disease. Fine Needle Aspiration Cytology(FNAC) proved to be useful in effective diagnosis obviating the need for biopsy.

Keywords: Cytomorphology, histopathology, Kikuchi-Fujimoto Disease. Lymphadenitis.

INTRODUCTION

Kikuchi-Fujimoto disease is a non-neoplastic condition characterized by lymphadenitis of necrotizing or reactive type with a mild predilection towards young females. It is an unusual condition with distinct clinical presentation often associated with autoimmune disorders especially Systemic Lupus Erythematosus (SLE), arthritis, polychondritis. Although it is a disease of unknown etiology, many theories have been postulated on the etiopathogenesis experiences. One the unique feature Kikuchi-Fujimoto disease [KFD] is the ‘self-limiting’ course with localized lymphadenopathy and non-fatal. Numerous agents had been found to be inciting factors like Ebstein-Barr virus[EBV], human papilloma virus [HPV6,8] and human immunodeficiency virus [HIV].Many Researchers opine that apoptotic induced cell destruction which is mediated by cytotoxic T lymphocytes and enhanced by histiocytes plays major role in the pathogenesis of the disease.

While KFD is more prevalent in Western countries, its incidence among the tropical countries, is rare entity,first reported in 2001. It presents with fever [low to high grade], skin rashes, joint pain and cervical lymphadenopathy with peripheral blood smear showing lymphocytosis and elevated ESR. Sometimes hepato-splenomegaly is revealed on sonology. It is not uncommon with idiopathic presentation with abrupt systemic signs and symptoms. Thus the condition mimics other clinical diseases e.g., lymphoma or extra-pulmonary tuberculosis lymphadenitis.

KFD always follows a dormant and benign course and many a times it is the associated autoimmune conditions brings the patient to the out-patient department [OPD].

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Thus clinical diagnosis of KFD is a challenging task and often warrant a pathologists for appropriate diagnosis. Lymph node biopsy for histopathological evaluation [HPE] by pathologist is the gold standard diagnostic procedure whereas culture and serology test plays role in exclusion of associated diseases like TB,SLE etc. Cytological diagnosis of KFD is vulnerable to subjective interpretation on microscopy.

The criteria for reporting KFD on cytology includes increased histiocytic population[crescentric shape], karyohectic debris, apoptotic bodies in background of inflammatory infiltrates of plasma cells, neutrophils and lymphocytes. However similar features are also encountered in TB [Non- mycobacterial TB] which often goes as necrotizing lymphadenitis advising biopsy, culture and serologic correlation. Hence it is evident that KFD cytological features shares several other conditions especially when presenting with non classical features as seen in this case report.

CASE REPORT

A 22 year old female farmer by profession from east coast of Pondicherry presented with cervical lymphadenopathy with low grade fever the for 2 months, joint pain and few rashes on extremities.

Right side lymph node was measuring 2x2cm with firm to rubbery consistency in antero-lateral triangle adjacent to hairline. No discharging sinus or punctum was noted. Moderate tenderness was noted on palpation. She was on antibiotic for 3 days which was ineffective. No clinical hepatosplenomegaly noted. Serological

Figure 1: FNAC smear shows sheets of lymphocytes with large mononuclear cells(arrow) (H&E, 40X)

Figure 2: FNAC smear shows sheets of lymphocytes in a background of debris(arrow). H&E, 40X.

Figure 3: Lymph node biopsy shows para cortical necrosis. (H&E, 10X).Inset: Histiocytosis with necrosis H&E40X
tests for Anti d-ANA and radiology proved to be non-contributory. Blood count showed lymphocytosis with elevated ESR. A clinical diagnosis of necrotizing/tuberculous lymphadenitis was made provisionally and referred for FNAC procedure. Cytology procedure was performed at our department as per standard protocol. Cytomorphology showed polymorphous population of cells composed of centroblasts, centrocytes admixed with numerous large atypical mononuclear cells in background of karyorrhctic debris suggesting non-neoplastic reactive lymph node lesion probably of histiocytic lymphadenitis. [Figures 1 & 2]. No granuloma was noted. Subsequent lymph node biopsy showed para-cortical necrosis with karyorrhctic and fibrin deposits, sheets of lymphoid cells with plasmacytoid, mononuclear cells corresponding to the atypical mononuclear cells on cytomorphology rendering the diagnosis of necrotizing histiocytic lymphadenitis – KFD [ Figure 3].The patient had a self-limiting course and recovered well after one month period.

**DISCUSSION**

KFD was first described by Dr. Masahiro Kikuchi in the year 1972 in Japan and established by Y. Fujimoto in 1980. It is due to exaggerated T cell mediated immune response in few functionally susceptible persons with unknown stimuli in young females.

While clinical interpretation is only provisional owing to diagnostic difficulties, excision biopsy for HPE is the mode of laboratory testing. While lymphoma and TB lymphadenitis warrants extensive treatment and therapeutic induced complications, KFD is self-limiting entity and non-fatal if diagnosed appropriately. FNAC has good reliability on lymph nodal lesions. While excision biopsy is an invasive time consuming procedure, FNAC is minimally invasive day care OPD procedure. Cytomorphology depends on the presence of histiocytic hyperplasia with reactive population of lymphocytes admixed with karyohexis with necrosis being the major clue to diagnosis. The challenging task for pathologist in tropical countries is exclusion of other similar diseases like TB lymphadenitis showing similar cytomorphology especially non-mycobacterial TB infection. First of its kind, the present case showed only reactive population without necrosis and corresponding biopsy confirming the diagnosis of KFD.

The reason attributed for the vague presentation is alteration in sequence of apoptosis and cytotoxic lymphocyte pathways. Another unique feature in the present case in non-association of autoimmune disease with serology negative tests making it a rare case report.

**CONCLUSION**

The present case study emphasizes on diagnostic utility of FNAC in appropriate diagnosis of KFD with unique presentation and non classical cytological features. Also KFD is known to occur even in the absence of associated autoimmune disorders and may present as non-necrotizing lymphadenitis as evident from this case report.

**CONFLICTS OF INTEREST**

None.

**REFERENCES**