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Case Report

Kikuchi-Fujimoto Disease in Patient with Cervical Lymphadenopathy: A Case Report

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ABSTRACT

Kikuchi-Fujimoto disease (Kikuchi Disease) is a self-limited and benign systemic lymphadenitis of unknown cause, originally described by Kikuchi and Fujimoto and coworkers in 1972. Although relatively uncommon, it is increasingly discussed in the medical literature. Clinical presentation typically includes adenopathy, particularly cervical with fever and flu-like symptoms. This constellation of symptoms, in the presence of a characteristic histiocytic necrotizing lymphadenitis, provides the clinicopathologic diagnosis. The immunopathogenesis of Kikuchi disease may lie in a hyperactive response to viral infection. It may mimic tuberculosis which is more prevalent in countries like India. Hence we should keep in mind differential diagnosis of Kikuchi's disease in patients with cervical lymphadenopathy.

Key words: Kikuchi disease, cervical lymphadenopathy, tubercular lymphadenopathy.

INTRODUCTION

Kikuchi-Fujimoto disease was first described independently by Kikuchi [1] and Fujimoto et al [2] as a group of patients presented with lymphadenitis showing focal reticulum cell hyperplasia, nuclear debris and phagocytosis and another with cervical sub acute necrotizing lymphadenitis, respectively. Hence, the entity is known as Kikuchi-Fujimotos disease or simply Kikuchi's disease (KD).

Since tubercular lymphadenopathy is the commonest cause for chronic cervical lymphadenopathy in developing and tropical country like India occasionally it is misdiagnosed, ignoring the other rare condition like KD if immunohistochemistry is not taken into consideration to differentiate. As a result the morbidity increases and cost of antitubercular treatment (ATT) in wrong diagnosed case is enormous.

The clinical and histological features suggest that this disorder represents a common pattern of response to a variety of etiologies. [3]

CASE HISTORY

23 yrs old female patient came to OPD with chief complaints of fever since 2 months and cervical lymhadenopathy since 2 months. No H/o wt loss, coughs, or contact with any tuberculosis patient. No H/o of any malignancy in family. She had been advised ATT on empirical basis by general practioner. Other systemic examination did not show any abnormality. Haematology showed mild pancytopaenia with normocytic red cell indices (Hb - 9.5 g/dl. MCV - 93.2 fl., Platelete count - 160

10^9/l) and a relative lymphopenia. Erythrocyte sedimentation rate (ESR) was 56 mm/h and C-reactive protein was 21. Excisional biopsy of a lymph node was done under local anesthesia. The histopathology demonstrated focal necrosis surrounded by karyorrhectic debris, histiocytes and plasmacytoid lymphocytes (Fig. 1). Kikuchi-Fujimoto disease (KFD) was diagnosed. The patient showed recovery after 30 days of supportive care.

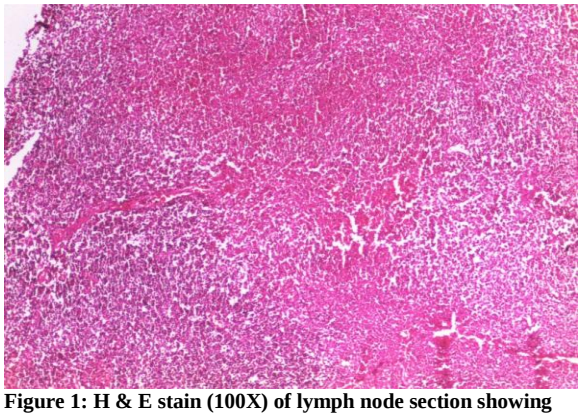
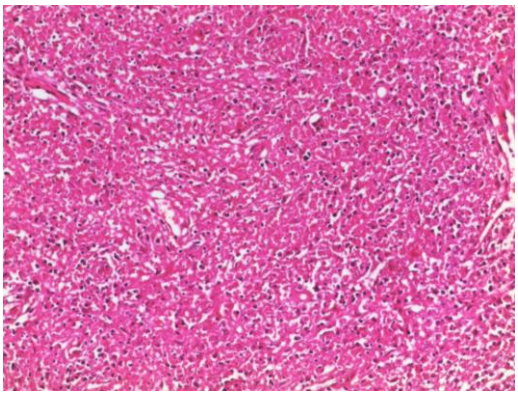
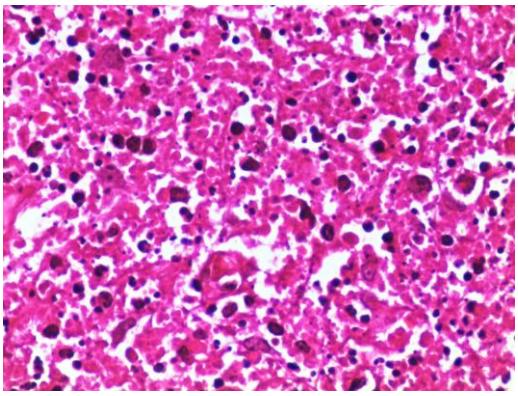
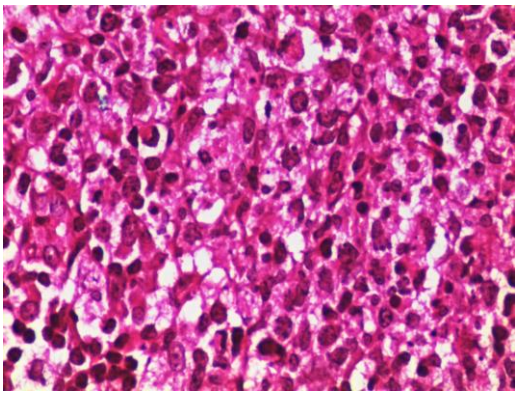
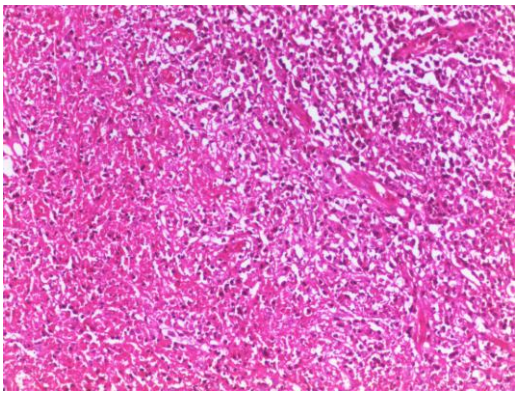


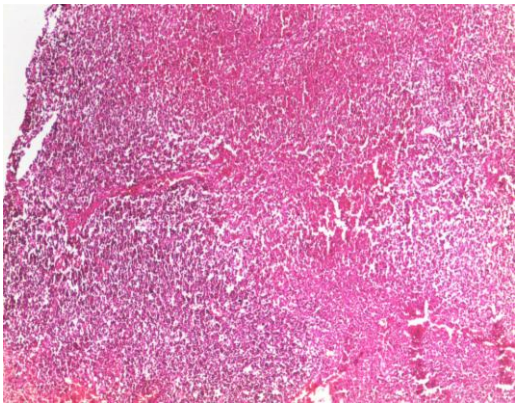
Figure 1: H & E stain (100X) of lymph node section showing Paracortical necrosis surrounded by karyorrhectic debris, histiocytes and plasmacytoid lymphocytes.

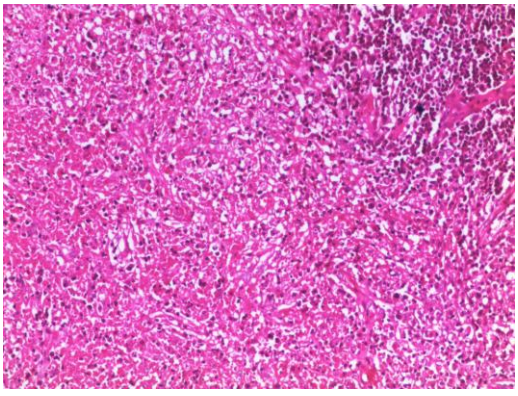












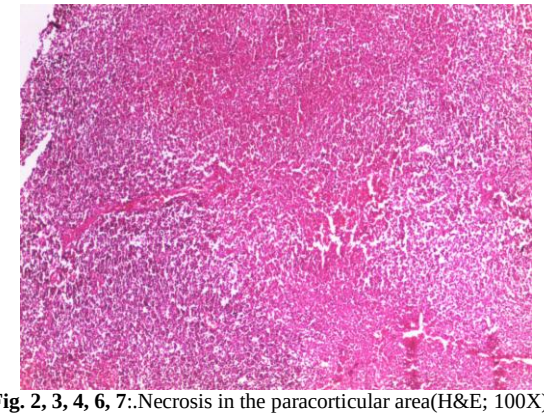


Fig. 2, 3, 4, 6, 7:.Necrosis in the paracorticular area(H&E; 100X)
Fig. 5: Prominent karyorrhexis and necrosis(H&E; 400X)
Fig 9: Histiocytes & lymphocytes at the periphery of the necrotic area.(H&E;400X)

DISCUSSION

Kikuchi-Fujimoto disease is also called histiocytic necrotizing lymphadenitis. It often presents as a painful cervical lymphadenopathy in young females. Unilateral and posterior cervical lymph nodes are the commonest to be involved. [4] The course of the disease is variable, but usually self-limiting. Less common manifestations are in the form of axillary and mesenteric lymphadenopathy, splenomegaly, parotid gland enlargement, arthralgias, myalgias, aseptic meningitis, bone marrow haemophagocytosis and interstitial lung disease [5-7]

The cutaneous lesions include erythematous macules, papules, plaques and nodules. [8] It is kept in the differential diagnoses of lymphadenopathy like infections including tubercular lymphadenitis, lymphoma, and other tumors. In Kikuchi disease, there is proliferation of T-cells, which, at some point, enter into the cycle of programmed cell death (apoptosis). [9] Initial studies hinted at Yersinis enterocolitica and toxoplasma gondii, however the absence of associated bacterial infection as well as the resolution of the illness without the use of antibiotics refute this hypothesis. [10] Viral infection has also been suspected, including Epstein-Barr virus (EBV), herpes virus 6, parainfluenza virus and cytomegalovirus. Serological tests including antibodies to EBV and other viruses have proved noncontributory. [10] Widespread activation of T-cell is also seen in some inflammatory joint diseases. The strongest link is with Systemic lupus erythematosis (SLE), although the exact nature of the association has not yet been established. [8] Since some patients of KD have latter developed SLE and since the necrotic lesion in the lymph node is similar, it has been suggested that necrotizing histiocytic lymphadenitis may be a forme firste of SLE. [11]

Patients with this Kikuchi Fujimoto disease may develop anemia, leucopenia, and atypical lymphocytosis and raised ESR. [12] Fine-needle aspiration cytology (FNAC) can be used to make cytologic diagnosis. Characteristic cytologic findings in KD include crescentic histiocytes, plasmacytoid monocytes, and extracellular debris. [13] The histology of KD is distinctive. There are patchy, irregular paracortical areas of coagulative necrosis without a polymorphonuclear leucocyte infiltration. The necrotic areas show prominent karyorrhectic debris, immunoblasts, histiocytes with characteristics C-shaped nuclei, some of which contain cellular debris, and plasmacytoid T-cells/monocytes. [10]

KD is self-limiting; the symptoms may spontaneously disappear in 1 - 6 months. Corticosteroids have shown good results. [14] The current evidence suggests the role of ciprofloxacin, chloroquin and hydroxychloroquin in this disease. [15]

Surgical care

Excisional lymph node biopsy for the purpose of confirming the diagnosis is the only surgery indicated in Kikuchi disease.

CONCLUSION

In patients with Kikuchi disease, diagnostic laboratory and radiologic test findings are nonspecific. Although results of fine-needle aspiration (FNA) may be suggestive. [16,17]

If FNAC is also not conclusive then IHC [immnunohistochemistry] is of great help. [18] In conclusion biopsy of all

suspicious cervical nodes is utmost important. It has further strengthened the histopathology in making the diagnosis more accurate in many clinical entities like sarcomas, carcinomas, and chronic inflammatory conditions. In our case, it helped in differentiating Kikuchi-Fujimotos disease from tubercular lymphadenopathy and guided to stop prophylactic anti tubercular treatment. Patient responded well to supportive treatment and recovered in 3 weeks.

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