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Kikuchi Fujimoto Disease – A Rare Case Report

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ABSTRACT :

Kikuchi-Fujimoto disease(KFD) is a rare and benign disease. It causes self-limited lymphadenopathy that typically affects young girls. KFD frequently manifests as weight loss, cervical lymphadenopathy and fever. In this disease, posterior cervical lymphadenopathy is the most commonest manifestation. The cause is idiopathic. Nonetheless, it is thought to be connected with autoimmune disorders and certain infectious agents. This disease is frequently misdiagnosed due to its rarity and vague symptoms. Immunohistochemical analyses and biopsies of the affected lymph nodes are crucial for obtaining a correct diagnosis.

Here, we present an interesting case of Kikuchi in twenty two years old female.

KEY WORDS : Kikuchi-Fujimoto, lymphadenopathy, Young females, biopsy, Immunohistochemistry.

INTRODUCTION :

"Histiocytic necrotising lymphadenitis," also known as Kikuchi-Fujimoto disease, is a benign, self-limiting systemic disorder that typically affects cervical lymph nodes of young patients and manifests clinically as fever and superficial lymphadenopathy.[1]

In 1972, Kikuchi and Fujimoto published the first report on it in Japan. [1] KFD has not been thoroughly studied because of its low incidence rate and vague clinical symptoms. [2] Although the pathogenesis of this disease is yet unknown, autoimmune responses and unknown infectious agents have been proposed as its primary causes. [3] This condition is most commonly found in the posterior cervical lymph nodes, and due to similar clinical characteristics, it is easily mistaken for other types of lymphadenitis. [4] This disease has become crucial for assessing current diagnostic practices and avoiding misdiagnosis and improper treatment. [5]

CASE REPORT :

A 22 year old female came to surgical OPD with chief complaints of swelling in right lateral neck region since 10 days. No complaints of pain, fever, trauma and no other associated co morbidities were seen.No history of similar complaints in the past. No significant family history noted.Vitals at the time of presentation were recorded – BP: 110/80mmHg, PR- 78/min, RR- 19/min, SPO2- 99%, temp- afebrile. Systemic examination was done and no abnormalities detected.

On examination of the swelling 3x2 cms and 4x3 cms nodes were present in the right side of neck at level 2 and 3 cervical region. Chest X-ray was normal and CT scan of the neck confirmed presence of several lymph nodes in right side of the neck with no other associated lesions.

FNAC was advised and performed on 21/10/2024 and reported as Granulomatous lymphadenitis. So to rule out Kochs etiology , excisional biospy was advised. All other blood investigations including complete blood picture, triple screening, renal profile were normal. Tuberculin skin test was also negative. Due to persistence of cervical lymphadenopathy despite treatment with analgesics and antibiotic therapy ,excisional lymph node biopsy was performed on 10/11/2024 and sent to the pathology department on 11/11/2024 for histopathological examination.Grossly,we received multiple grey white soft tissue bits,largest measuring 0.5x0.5 cm and smallest measuring 0.2x0.2cm.Histopathological examination show fragments of lymph node tissue with partially distorted architecture displaying necrotic areas composed of eosinophilic, granular material, karyorrhectic debris and interspersed lymphocytes and histiocytes .These areas are surrounded by histiocytic cells and plasmacytoid dendritic cells. There is an expansion of small lymphocytes admixed with few immunoblasts. Residual primary and secondary follicles are present. So the impression was given as “Necrotising lymphadenitis ,favouring Kikuchi disease” and advised Immunohistochemistry panel for further confirmation.

Immunohistochemistry was done outside, revealed MPO and CD68 diffuse positivity and other lymphoma markers turned out to be negative, thus favoured Kikuchi –Fujimoto disease.

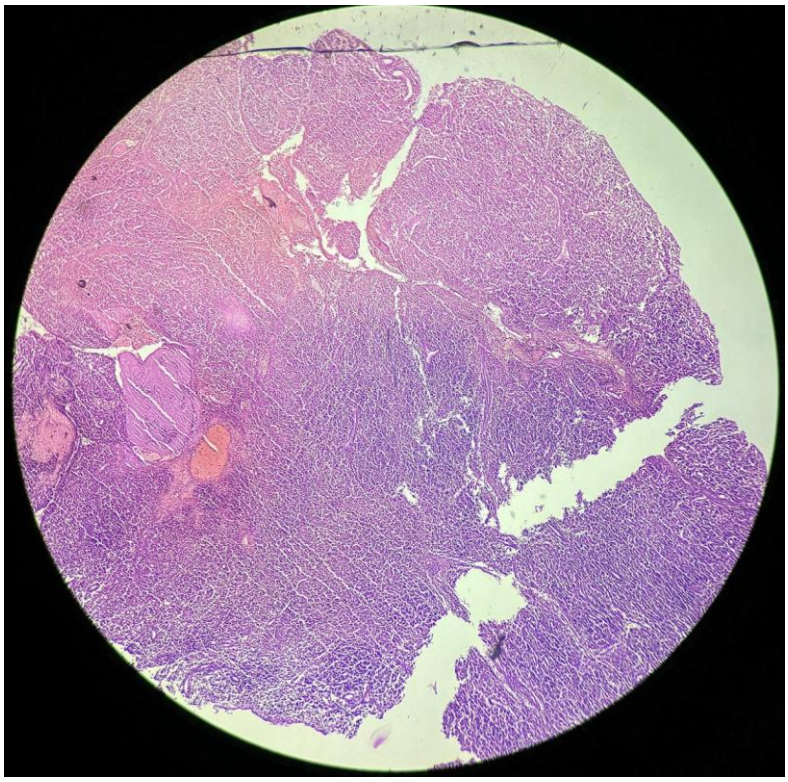

Fig 1: Low power view,H&E,showing lymphnode tissue with distorted architecture.

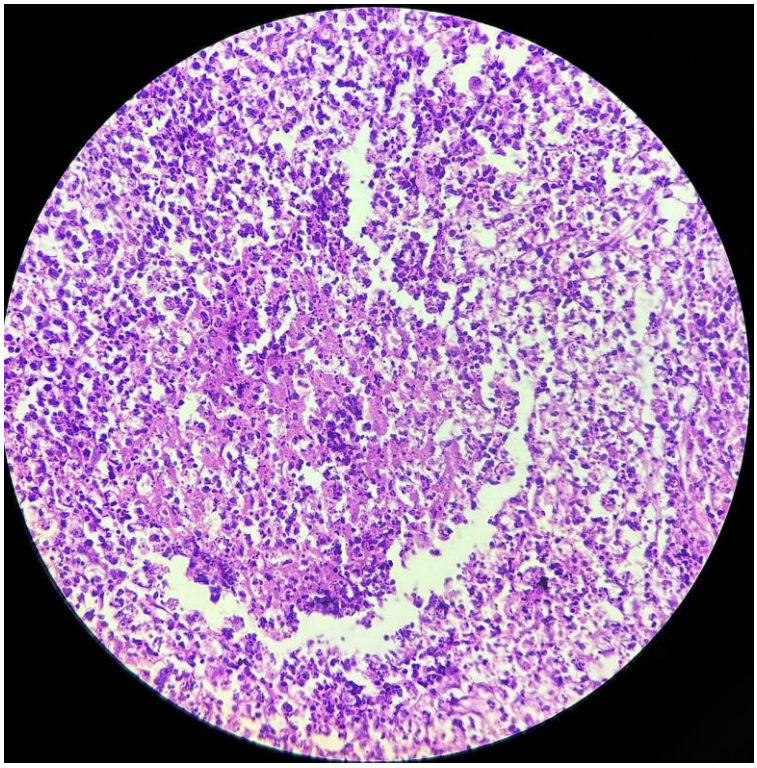

Fig 2: High power view,H&E,showing necrotic areas composed of eosinophilic granular material, karryorhectic debris and interspersed lymphocytes and histiocytes.

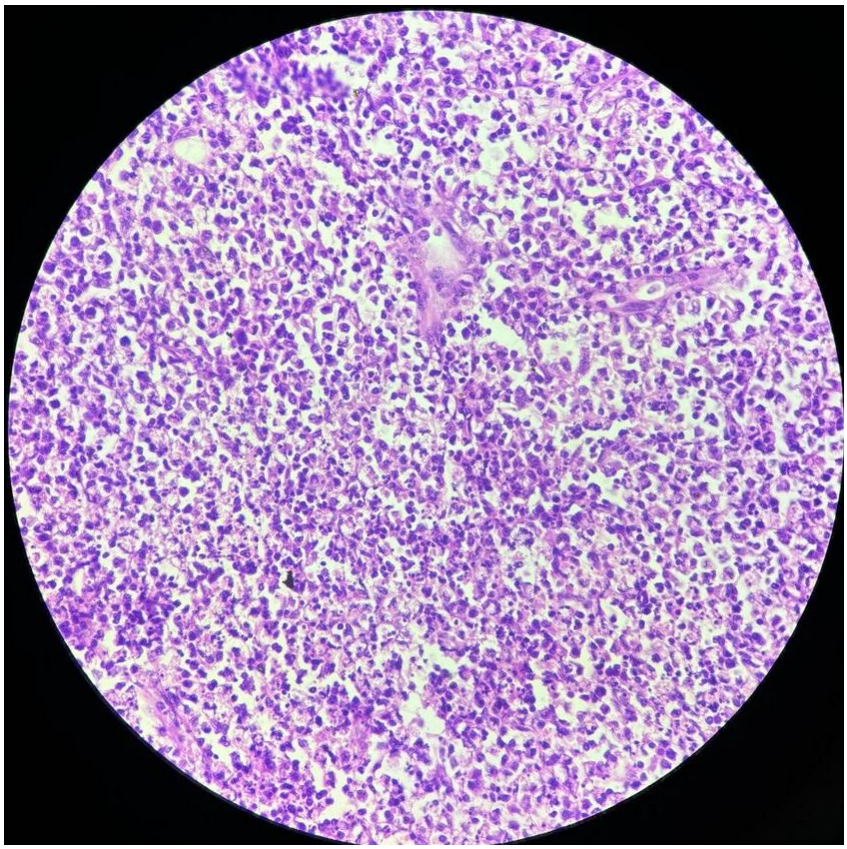

Fig 3: High power view,H&E, showing karryorhectic debris and interspersed lymphocytes and histiocytes.

DISCUSSION :

Histiocytic necrotising lymphadenitis, often known as Kikuchi-Fujimoto disease, is a rare idiopathic cause of lymphadenopathy that was separately reported by Kikuchi and Fujimoto et al. in 1972. [1] It is challenging to distinguish it from other lymphadenopathy causes such as lymphoma, Tuberculosis and mononucleosis. [1] The prevalence of this disease is higher in western and Asian nations. According to some experts, Kikuchi-Fujimoto disease is linked to HLA class II alleles (DPA 101 and DPB 102), which are more common in Asian people. [4]

Typically manifests in young adults, with a slight female preponderance and an average age of onset is 21 years. [5] Clinical manifestations and pathological features are the two most crucial aspects in the diagnosis of this disease.[6] Most common clinical presentations are fever and cervical lymphadenopathy in previously young adults, as in our case. Additionally, it may be associated with skin rashes and sore throat. A few reported fatal cases have been reported where fulminant hepatitis, necrotizing myocarditis, organ transplantation were associated with fever and lymphadenitis. [7] Laboratory investigations are fairly non-specific. Most common finding is leukopenia with mild lymphocytosis and occasional atypical lymphocytes. [8] However majority of the patients have a normal complete blood cell count. The patient in our study has normal complete blood picture. Pathological analysis of lymphnode biopsy is frequently performed, which generally show extensive areas of necrosis and histiocytosis in the cortical and paracortical regions of the lymphnode. Other histological features such as, clusters of mononuclear cells with scattered nuclear debris are indistinguishable from lymphoma. So immunohistochemical studies help in differentiating these tumors. [9]

CONCLUSION :

Kikuchi-Fujimoto disease is a benign and rare disease that usually occurs in young females. Clinical manifestation varies and can be mimic other diseases. Pathological biopsies and immunohistochemistry are the basis for the diagnosis. In young females who develop cervical lymphadenopathy for a short period of time , this diagnosis should be considered. To evaluate these patients for the onset of autoimmune disease or its recurrence, long-term follow-up is necessary.

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