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KIKUCHI FUJIMOTO DISEASE –NECROTISING CERVICAL LYMPHADENITIS

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Abstract: *Kikuchi- Fujimoto disease is a rare benign self-limited disease of unclear etiology. It is important to recognize and diagnose this entity as it closely mimics and is often mistakenly clinically diagnosed as lymphoma. Case report: - We report a case of young male of 24 years age with fever and tender cervical lymphadenopathy. FNAC suggested a diagnosis of reactive lymphadenitis and patient put on broad spectrum antibiotics. As the patient showed no response the biopsy of the cervical lymph node was done and microscopy suggested the diagnosis of Kikuchi disease. Conclusion: - Kikuchi Fujimoto disease is a rare self-limited disease, more common in females .Our case is again a rare case of Kikuchi's disease reported in a male patient. Clinician and pathologist should be aware of this entity and keeping it as a close differential diagnosis in appropriate cases will minimize the unnecessary treatment and its side effect.*

Key Words: *Histiocytic, Kikuchi-Fujimoto Disease, Necrotising Lymphadenitis*

Introduction

Kikuchi- Fujimoto disease is a rare benign self-limited disease, more common in females as compared to males presenting as painless cervical lymphadenopathy. We describe a case of Kikuchi Fujimoto disease presenting as an unusual case of cervical lymphadenopathy in a young male. The histopathology confirm the diagnosis of Kikuchi Fujimoto disease without any difficulty but this case stresses the importance of prompt biopsy of isolated cervical lymphadenopathy in young male as well as female patients. Early and correct diagnosis of this entity minimizes unnecessary treatment and their side effects.

Case History

A twenty four year old male presented to the medical outpatient department with complaints of fever and cervical lymphadenopathy. Initially only cervical lymphadenopathy was noticed by the patient which he reported showed an increase in size over a period of four weeks. Axillary lymphadenopathy was noticed by the physician on general physical examination. No direct history of cough, breathlessness or any past history of hospitalization for any other medical problem was given by the patient. There was no history of weight loss.

On examination left cervical and axillary lymphadenopathy confirmed, measuring 1 to 2cm in diameter. Systemic examination was normal. On conducting laboratory investigations, he was found to be anemic with a hemoglobin of 7 gm%, total leukocyte count 5.4×10^9/L, absolute lymphocyte count 2.3×10^9/L, ESR 45mm/hr, negative blood and throat culture, chest radiograph was normal, contrast CT scan of the neck showed non uniform enhancement of the lymph node

with central low attenuation necrotic areas. Tuberculosis and Lymphoma was suspected as a first differential diagnosis.

Fine needle aspiration cytology of the cervical lymph node showed polymorphic population comprising of reactive lymphoid cells giving an impression of reactive lymphadenopathy. No evidence of granulomatous pathology seen and the ziehl neelsen stain for acid fast bacilli was also negative. An excision biopsy of the largest cervical lymph node was performed under local anaesthesia, tissue fixed in 10% aqueous formalin and subjected to histopathological examination. Microscopic examination of the lymph node show an intact capsule variably altered architecture along with characteristic cortical foci of necrosis with an appreciable number of karyorrhectic nuclei surrounded by a polymorphous population of histiocytes, small and large lymphocytes with paucity of plasma cells and absence of neutrophils. No granuloma was there. A part of the section shows residual lymphoid follicles and variable follicular hyperplasia. A diagnosis of Kikuchi disease was made and patient kept under follow up. Patient remained well and showed complete recovery three months later.

Discussion

Necrotizing lymphadenitis (Kikuchi's lymphadenitis), Kikuchi-Fujimoto disease is seen most frequently in Japan and other Asian countries , but also occurs in other countries including United States and Western Europe and more common in females as compared to males, and that too in younger age group with a persistent painless cervical lymphadenopathy accompanied by fever[1] . This disease was first described in 1972 independently by Kikuchi [2] and Fujimoto[3]. Its true incidence is .unknown [4]. Female to male ratio varies between 4:1[5].Kikuchi disease affect a wide range from 20 months to 75 years, with a mean patient age in the third decade[6.] Involvement of nodal areas other than cervical lymph nodes has been discernible including axillary, thoracic, abdominal and pelvic. The pathogenesis is unclear but a viral or postviral hyper immune reaction has been suggested as a possible mechanism[4]. Special studies have shown that necrosis is an expression of cytotoxic lymphocyte mediated apoptotic cell death. Recent studies have shown that diagnosis can be made or atleast suspected from presence of phagocytic histiocytes with peripherally placed nuclei and plasmacytoid monocytes[1]. The disease is generally benign and self-limited usually resolves in several weeks to months. The disease has a recurrence rate of 3 to 4%[7]. The most important differential diagnosis is with Malignant Lymphoma with secondary necrosis[1] Morphological features that favor Kikuchi Fujimoto disease over lymphoma are the patchy non expansile distribution of the lesion with incomplete architectural effacement with patent sinuses, abundance of karyorrhectic debris, presence of admixed medium-sized cells with round nuclei (plasmacytoid dendritic cells), numerous reactive histiocytes without a starry-sky pattern , relatively low mitotic rate , and presence of intervening areas with reactive appearance [8]. William primrose *et al* reported a case of Kikuchi's presenting with posterior triangle lymphadenopathy. On examination clinician confirmed 2cm diameter painless rubbery lymph nodes. Systemic examination and laboratory investigation were normal. In this case also the clinician suspicion was of lymphoma but the histopathology gave a diagnosis of Kikuchi-Fujimoto disease[5]. Sudhakar *et al* in their study also reported a case of young female of South Indian origin with a short history of cervical lymphadenopathy and fever. Clinical examination revealed bilateral tender cervical lymphadenopathy. FNAC suggested a diagnosis of reactive lymphadenitis. But histopathology confirmed the case to be of Kikuchi Fujimoto disease[9]. Most of the references report that Kikuchi-Fujimoto disease is more common in females with male to female ratio (1:4). But a recent review article reports that the female predominance is overemphasized but the actual male to female ratio is 1:1[10.] The cause of this particular form of lymphadenitis remains uncertain but may be multifactorial. To date, all reported cases have had a benign course but usually resolving within a period of few months, but the recurrence rate of 3-13 % has been reported. Kikuchi Fujimoto disease does not carry the increased risk of Lymphoma but few cases may progress to autoimmune disease and this is the reason why patients should be kept under regular clinical follow up[10].

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