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Case Report

Kikuchi - Fujimoto Disease - A Rare Case of Cervical Lymphadenopathy

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ABSTRACT: A rare case of Kikuchi-Fujimoto disease presenting as a slow-growing cervical lymphadenopathy in a 32 year old male patient has been reported with discussion on the problems in diagnosing such a case.

Key Words: Necrotizing lymphadenitis; Kikuchi- Fujimoto disease; Cervical lymphadenopathy.

INTRODUCTION: Kikuchi-Fujimoto disease (KFD; histiocytic necrotizing lymphadenitis) is a rare, benign, and self-limited syndrome characterized by regional tender lymphadenopathy, usually accompanied by mild fever and night sweats. It is an extremely rare disease predominant among Japanese. It is mainly a disease of the young adults (20- 30 years) with female predominance. It was first reported almost simultaneously by Kikuchi and by Fujimoto and associates in 1972 as a lymphadenitis with focal proliferation of reticular cells accompanied by numerous histiocytes and extensive nuclear debris[1].

CASE REPORT: A 32 year old male presented with multiple, slow growing, mildly tender, mobile, firm lymph nodes involving the upper, middle and lower deep cervical region (Fig.1). The patient complained of gradually increasing swelling on the right side of the neck with moderate degree, midday (12 noon- 2 pm) fever which was associated with night sweat, intense itching over the body and multiple joint pain (more in upper extremity than lower extremity) without any signs of loss of appetite, malaise or loss of weight. Systemic examination did not reveal any axillary/ inguinal lymphadenopathy or splenomegaly. Chest X-ray showed upper mediastinal widening (Fig.2) and Ultra sound abdoven showed hepatic enlargement. Other routine tests were within normal limits. As the FNAC was inconclusive, incisional biopsy was planned. The first two biopsies were inconclusive and the 3rd one suggested the diagnosis of Kikuchi-Fujimoto disease characterized by areas of eosinophilic necrosis, leaving behind scanty lymphoid tissue at periphery with partial effacement of architecture, attenuated follicles, prominent histiocytes, eosinophilic infiltrate (Figs.3 & 4). No evidence of granuloma was seen in the specimen. ZN stain did not reveal any acid fast bacillus.

He was treated with 36 mg/ day of methylprednisolone in divided doses with gradual tapering of dose over 1½ months, NSAIDs and antibiotics for three weeks. The patient showed improvement after one months' treatment with dramatic reduction in size of the swellings. The neck nodes disappeared after two months. Intermittent fever still persisted. Joint pain significantly reduced itching is negligible. The patient was kept under surveillance without any medication. After 6 months of regular follow-up, no sign of recurrence could be detected.

DISCUSSION: Kikuchi- Fujimoto disease is an enigmatic, benign and self-limited syndrome characterized by regional lymphadenopathy with tenderness, predominantly in the cervical region, usually accompanied by mild fever and night sweats. Initially described in Japan, it was first reported in 1972 almost simultaneously by Kikuchi[2] and Fujimoto[3] et al as a lymphadenitis with focal proliferation of

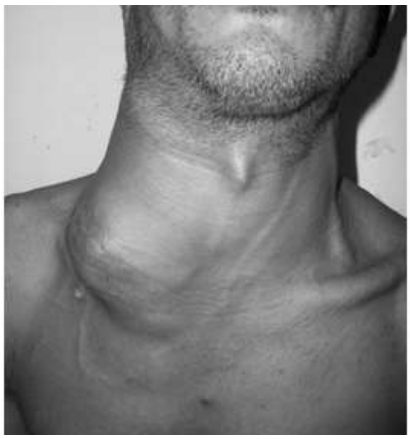

Figure 1: Clinical photograph of the patient Kikuchi-Fujimoto disease showing right sided neck swelling and scar of the biopsy procedure.

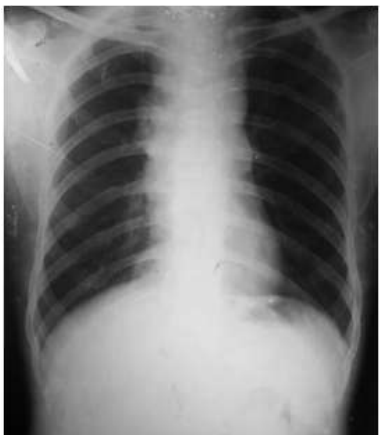

Figure 2: Chest radiograph shows mediastinal widening.

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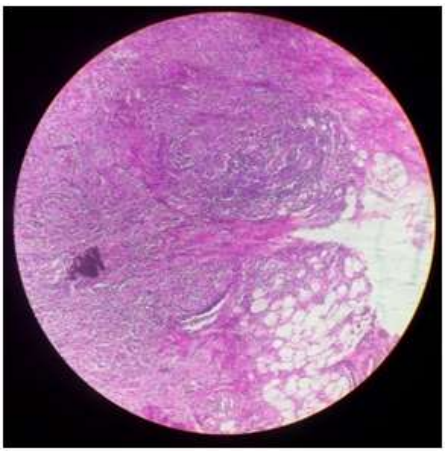

Figure 3: Histopathological section of a cervical lymph node showing areas of eosinophilic necrosis, Attenuated lymphoid follicles with partial effacement of architecture, prominent histiocytes and eosinophilic infiltrate (H&E x 10)

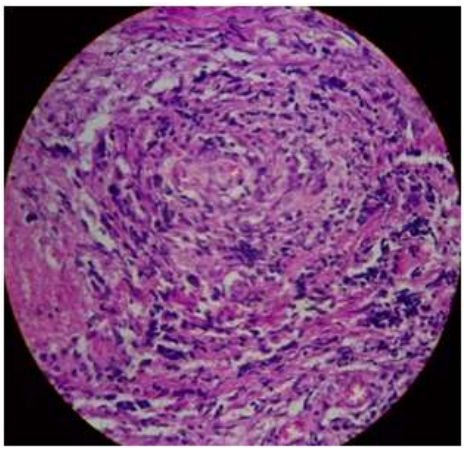

Figure 4: Histopathological section of a cervical lymph node showing eosinophilic infiltrate, areas of necrosis, attenuated follicles, prominent histiocytes (H&E x 40)

reticular cells accompanied by numerous histiocytes and extensive nuclear debris[4]. The age range is wide (11–80 years of age) but the majority of patients are under 30 years of age. Clinically cervical lymph nodes are the principal site of disease and are reported in 70–98% of patients[5]. Involvement of most other nodal areas has been described including axillary, thoracic, abdominal and pelvic. Splenomegaly has also been described[5]. The patient, in this report, was 30 year old male with right sided cervical lymphadenopathy with mediastinal involvement and hepatomegaly without any features of axillary, abdominal, inguinal lymphadenopathy or splenomegaly. Fever is the primary symptom in 30-50% of cases. Less common manifestations include weight loss, chills, skin rash, gastrointestinal symptoms and night sweats[5]. These features are corroborating with our case except absence of any gastrointestinal symptoms. The results of laboratory investigations are often normal. The pathogenesis is unclear but a viral or post viral hyperimmune reaction has been suggested as a possible mechanism. The viral agents implicated based on virological teste are Epstein Barr Virus (EBV), Human Herpes Virus(HHV)[6,7,8], Parvovirus B19[9,10]. Autoimmune studies are usually negative. However, cases associated with systemic lupus erythematosus (SLE) and Hashimoto disease have been reported. Complete spontaneous recovery is usual[5] Kim and Fulcher each described a case of Kikuchi disease with diffuse homogeneously enhancing cervical lymphadenopathy. Intraparotid, supraclavicular and mediastinal adenopathy were also present in one case. The CT features of homogeneously enhancing nodes without significant nodal necrosis make radiological differentiation from lymphoma impossible. Two cases reported in literature with MRI and contrast enhanced CT. One case had similar CT findings to the other reported cases whilst the other showed inhomogeneous enhancement with extensive nodal necrosis[8]. The pathological features may mimic lymphoma in some cases owing to the variable degree of microscopic necrosis found within the lymph nodes as well as the atypical appearance of the histiocytes, which may be confused with the small and large cleaved follicular cells seen in some cases of lymphoma. However, the absence of granulocytes and plasma cells usually allow the differentiation of Kikuchi disease from lymphoma or lymphadenitis caused by bacteria or viruses. The morphological distinction from SLE may be problematic in some cases[4] Spontaneous complete resolution of symptoms usually occurs within 4 months. CONCLUSION: The Kikuchi-Fujimoto disease presents with the features similar to multiple array of diseases and posses a unique diagnostic dilemma. Histopathological examination of lymph node biopsy is the diagnostic procedure of choice and sometimes need multiple biopsies. Conservative treatment with methylprednisolone, NSAIDs and antibiotics usually controls the symptoms.

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