KIKUCHUI- FUJIMOTO DISEASE

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INTRODUCTION

Kikuchi's disease first described in 1972 by Kikuchi and Fujimoto et al usually manifests as a localized cervical adenopathy, primarily in the posterior neck of young women [1]. Its course is benign, but can be misdiagnosed as lymphoma, both clinically and pathologically [2].

CASE REPORT

A 29-year-old woman presented with a history of fever and multiple swellings over both sides of neck for last one month. She was evaluated initially in a different hospital and considering the high local prevelance of tuberculosis underwent excisional lymph node biopsy of right cervical lymph nodes which revealed chronic nonspecific inflammation. However postoperatively on third day she developed swelling over the biopsy site with purulent discharge and high grade fever accompanied by rigors and chills. Mantoux test was negative as were tests for human immunodeficiency virus and brucellosis. Her angiotensin-converting enzyme level was normal. Her antinuclear antibody (ANA) screen was negative. Pus swab from the biopsy site revealed staphylococcus aureus sensitive to Flucloxacillin. Findings on chest x-ray were interpreted as normal. Patient started improving within a week.The swelling subsided and purulent discharge stopped. However she continued to have low grade evening fever with night sweats. After two weeks an excisional biopsy of cervical lymph node from the left side was done. Histopathology report revealed partial effacement of the lymph node architecture with focal cortical and paracortical areas of karyorrhexis and karyolysis. Many histiocytic cells with peripherally placed crescentic nuclei were also seen. Ziehl Neelsen stain and Periodic acid Schiff stains failed to demonstrate acid fast bacilli and fungi respectively. Based on these findings a diagnosis of Kikuchi Fujimotos disease was formulated (Fig. 1 & 2). Patient and her husband were reassured of the benign nature of the disease and patient was discharged with only ferrous gluconate to treat her iron deficiency anemia. Patient was followed up and she became afebrile in six weeks with complete resolution of cervical lymphadenopathy over three months. She has been followed up for a year and a half now and she remains asymptomatic without any clinical evidence of an associated connective tissue disease uptill now.

DISCUSSION

Kikuchi,s disease usually manifests as a localized cervical lymphadenopathy, primarily in young women. The female-to-male ratio in Kikuchi's disease is approximately 4:1 and most patients are in their late 20s or early 30s (mean age: 30 years) as was in this case [3]. Although the large majority of Kikuchi's disease cases have been seen in Japan, where it was initially described, many other countries have since added cases to the literature including Saudi Arabia [1]. It has been suggested that most patients with this disease are Asian, but 68 of Dorfman and Berry's 108 patients (63.0%) were white [3].

The etiology of Kikuchi's disease is controversial. Several possible etiologies have been postulated. Among the infectious organisms that have been proposed are Yersinia enterocolitica, cytomegalovirus (CMV), human herpesvirus (HHV), varicella-zoster virus, parainfluenza virus, and Epstein-Barr virus (EBV). However, studies thus far have failed to demonstrate a relationship between Kikuchi's disease and either of these [3, 4]. Likewise, Martinez-Vazquez et al evaluated seven lymph node specimens by

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Fig. 1: scanner view showing necrotizing change in the cortical and paracortical region of lymph node (H*E*40)

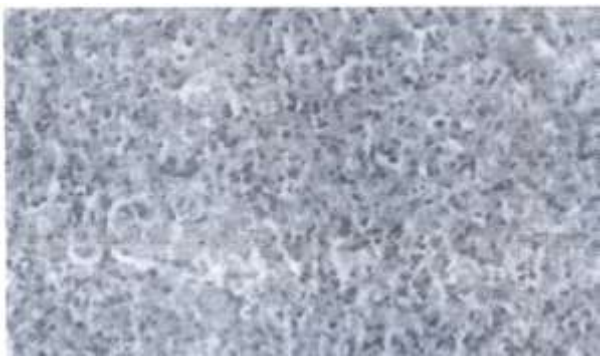

Fig. 2: High power view shows areas of Karyrrexis / pykonsis in Lymph node (H*E*200)

polymerase chain reaction for viral DNA (specifically, herpes simplex virus types 1 and 2, varicella-zoster virus, CMV, HHV6, HHV8, and EBV) and found no evidence implicating these pathogens[5]. Moreover, toxoplasmosis, autoimmune aetiology have also thught to be factors. The possible association between Kikuchi's disease and SLE is important because Kikuchi' s disease might be the initial diagnosis in patients who go on to develop SLE. It has been suggested that Kikuchi's disease might represent a self-limited SLE-like autoimmune disorder [6, 7]. Therefore, it is recommended that patients with Kikuchi's disease be monitored long term for the development of SLE [3].

Most patients have cervical lymphadenopathy of modest dimensions. Other affected sites include the axillary (14%) and supraclavicular (12%) nodal chains. (5) Additional signs and symptoms include fever, hepatosplenomegaly, headache, anorexia, nausea, vomiting, skin lesions, and constitutional disturbances (e.g., night sweats, weight loss, and malaise). The patient in this study had fever, night sweats and painless cervical lymphadenopathy. Lupus lymphad-enitis and malignant lymphoma can be radiologically similar to Kikuchi's disease, and they should be ruled out.

Most patients with Kikuchi's disease also have elevated erythrocyte sedimentation rates and C-reactive protein levels [3]. The patient of this study had elevated ESR but had neutophilia which was due to the superadded abscess post biopsy. Hsueh et al have advocated that fine-needle aspiration of the lymph node alone should be sufficient for a diagnosis [8], but most other authors rely on a formal biopsy for tissue diagnosis and same was done in this patient. Histologic analysis of affected lymph nodes generally reveals patchy necrosis, primarily in the paracortical area, and numerous adjacent crescentic histiocytes, lymphocytes, macrophages, and immunoblasts (predominantly T cell). The histiocytes and macrophages contain phagocytized nuclear debris from degenerated lymphocytes. Granulomas are absent. These histologic features, in conjunction with an absence or paucity of neutrophils, are consistent findings in Kikuchi's disease. In 1995, Kuo studied histopathologic changes in 79 patients with Kikuchi's disease and proposed three histopathologic types: proliferative, necrotizing, and xanthomatous [9].

Kikuchi's disease itself usually does not require treatment, although Jang et al suggested that patients with advanced disease might benefit from systemic prednisone to speed resolution.Treatment is indicated, of course, for patients whose disease is complicated by other factors. For examlpe acetaminophen or a nonsteroidal anti-inflammatory drug might be necessary to treat pyrexia and/or lymphadenopathic pain. Symptoms of Kikuchi's disease tend to spontaneously disappear in a matter of weeks to 6 months; in some rare cases, symptoms have persisted for a longer period. A recurrence rate of 3.3% has been reported [9]. Two deaths associated with Kikuchi's disease have been reported in the literature.

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