Case Report

Our patient Mrs. Sumona aged 21 years, newly married Bangladeshi girl presented to us in December 2010 with a swelling in her neck for 3 months with mild pain. She complained severe throat pain with difficulty in swallowing for 5 to 6 days. She also complained of pain in her both knee joints and runny nose. On examination, tonsils were swollen, congested, reddened, with slight foetor oris and some dehydration. Nose shows oedematous and pale looking nasal mucosa with bilateral hypertrophied inferior turbinate. Neck shows enlarged lymph nodes in the posterior triangle which were firm in consistency and less mobile, slightly tender and multiple in number. Skin over the swelling was free. The jugulo digastric lymph nodes were also found to be enlarged and tender on both sides. Neck examination also showed previous scar marks above and near the present swelling due to previous excision biopsy. The axillary and inguinal lymph nodes, liver and spleen were not palpable. Indirect laryngoscopy was done later on and no abnormality was detected. There was no rash seen in her body at that time. She was provisionally diagnosed as a case of recurrent tonsillitis with lymphadenopathy in her left posterior triangle with allergic rhinitis.

She was prescribed non steroidal anti inflammatory analgesic (ibuprofen 300 mg 3 times daily for 7 days in full stomach and oral antibiotic Levofloxacin 500 mg once daily for 14 days for her lymphnodes swelling and throat pain.). She was also prescribed an antihistamine for 14 days and a nasal spray containing xylometazoline and sodium chromoglycate for 21 days.

Her Hb% was 12.1 gm/dl & ESR 70 mm in 1st hour. Tuberculin test was reported as doubtful. X-ray chest & USG of abdomen were normal. RA test, serum LDH, ANA, anti-ds-DNA, IgG, IgM and IgA, and CT scan of the brain were normal. Excision biopsy of cervical lymph node was done and sent for histopathology. A diagnosis of necrotizing lymphadenitis (Kikuchi Lymphadenitis) was made by the histopathologist and the patient was diagnosed as a case of recurrent KFD (Kikuchi-Fujimoto disease).

Discussion

Kikuchi disease, also called histiocytic necrotizing lymphadenitis or Kikuchi-Fujimoto disease is an uncommon, idiopathic, generally self-limited cause of lymphadenitis. Kikuchi first described the disease in 1972 in Japan. Fujimoto and colleagues independently described Kikuchi disease in the same year. The most common clinical manifestation of Kikuchi disease is cervical lymphadenopathy, with or without systemic signs and symptoms. Clinically and histologically, the disease can be mistaken for lymphoma or systemic lupus erythematosus. Kikuchi disease typically affects young adults (mean age, 20-30 y) & almost always runs a benign course and resolves in several weeks to months. Disease recurrence is unusual, and fatalities are rare, although they have been reported. Several authors have reported an association between Kikuchi disease and SLE. Kikuchi disease has been diagnosed before, during, and after a diagnosis of SLE was made in the same patient. Additionally, the histologic appearance of lymph nodes in patients with Kikuchi disease is similar to that of lymph nodes in patients with SLE lymphadenitis. The association of Kikuchi disease with SLE, if any, remains unclear.

Although uncommon, Kikuchi disease has been reported throughout the world and in all races. Most cases have been reported from East Asia, with fewer cases from Europe and North America. The course of Kikuchi disease is generally benign and self-limited. Lymphadenopathy most often resolves over several weeks to 6 months, although the disease occasionally persists longer. The disease recurs in about 3% of cases.

Cervical nodes are affected in about 80% of cases. Posterior cervical nodes are frequently involved (65-70% of cases). Lymphadenopathy is isolated to a single location in 83% of cases, but multiple chains may be involved. Cases of generalized adenopathy involving axillary, inguinal, and mesenteric nodes are unusual. A flulike prodrome with fever is present in 50% of cases.
The incidence of skin involvement varies from 5-30% and includes maculopapular lesions, morbilliform rash, nodules, urticaria, and malar rash, which may resemble that of SLE. Skin lesions resolve in a few weeks to months. Hepatosplenomegaly is not uncommon. Neurologic involvement is rare but has included conditions such as aseptic meningitis, acute cerebellar ataxia, and encephalitis. Widespread involvement of multiple organ systems in Kikuchi disease has been described in solid-organ transplant patients.4,5

On histopathology the numerous atypical monocytes and T-cell immunoblasts observed in Kikuchi disease may lead to an erroneous diagnosis of lymphoma, especially high-grade non-Hodgkin’s lymphoma. Incomplete architectural effacement with patent sinuses, presence of numerous reactive histiocytes, relatively low mitotic rates & absence of Reed-Sternberg cells may help prevent its misdiagnosis as malignant lymphoma. Positive immunostaining results by monoclonal antibody Ki-M1P are seen in Kikuchi disease but not in malignant lymphoma. Kikuchi disease and SLE have similar histopathologic appearances. Distinguishing the two entities can be difficult.6

Treatment of Kikuchi disease is generally supportive. Nonsteroidal anti-inflammatory drugs (NSAIDs) may be used to alleviate lymph node tenderness and fever. The use of corticosteroids, such as prednisone, has been recommended in severe extranodal or generalized Kikuchi disease. Indications for corticosteroid use include the following: neurologic involvement, aseptic meningitis, cerebellar ataxia, hepatic involvement – elevated LDH level and severe lupuslike syndrome – Positive ANA titers. Jang and colleagues recommended expanding the indications for corticosteroid use to less severe disease. They administered prednisone when patients had prolonged fever and annoying symptoms lasting more than 2 weeks despite NSAID therapy, as well as for recurrent disease and for patients who desired a faster return to work. Immunosuppressants have been recommended as an adjunct to corticosteroids in severe, life-threatening disease.1,3,5,6 Kikuchi disease is generally a self-limited disease with a favorable prognosis. Lymphadenopathy usually resolves within 1-6 months after onset, although it may persist longer. About 3% of patients experience recurrence.6

References