Fever and Cervical Lymphadenopathy in a Young Female; Thinking beyond Tuberculosis

Sir,

A 28-year-old female presented with bilateral multiple cervical lymphadenopathy and fever for 6 weeks [Figure 1]. There was cervical lymphadenopathy (bilateral, multiple, mobile, nontender and largest being 2 cm × 2.5 cm), thyromegally and hepatomegaly of 5 cm. Investigations revealed leukocytosis with relative lymphocytosis and raised inflammatory markers. Liver enzymes were elevated up to 200-300 IU/L. Markers for hepatitis B virus, hepatitis C virus, HIV, TORCH profile and Epstein — Barr virus were negative. Thyroid function test revealed T3-0.7 nmol/L, T4-64 nmol/L, thyroid stimulating hormone 64 and anti-thyroid peroxidase antibody-576 KIU (normal <35). Ultrasonography of neck revealed features of chronic thyroiditis. Excision biopsy of one of the lymphnode showed histiocytic necrotizing lymphadenitis of Kikuchi's disease [Figure 2]. The patient was treated with indomethacin 75 mg bid. As the lymphadenopathy did not resolve after 2 weeks, we added oral prednisolone (0.5 mg/kg) for a period of 4 weeks, which was tapered over the next 4 weeks. The patient is fine even after 1-year.

Only about 20 cases from India and some hundreds of Kikuchi's disease from all over the world has been reported since it was first described by Kikuchi in Japanese females in 1972. The exact cause is unknown. Viral and postviral etiology have been proposed. Epstein Barr virus has been most consistently studied.[1] There have also been reports of a possible link with autoimmune disorders like systemic lupus erythematosus (SLE) and autoimmune thyroiditis.[2] The

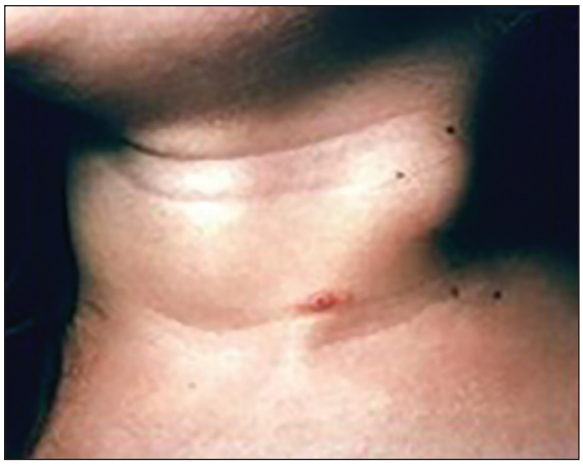

Figure 1: Bilateral neck swelling

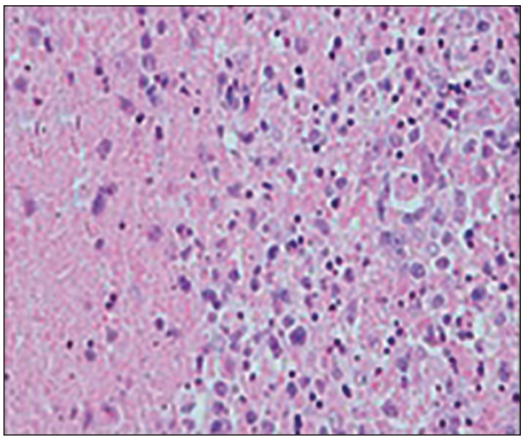

Figure 2: (H and E, ×100) showing histiocytes, lymphocytes and few neutrophils

classical clinical features of sub-acute fever with unilateral, discrete, and tender regional lymphadenopathy have often been confused with tuberculosis, SLE and lymphoma. Extra nodal involvement may occur in skin, bone marrow, myocardium and central nervous system.[3] Systemic symptoms such as anorexia, sore throat, arthralgia, weight loss, and night sweat have been reported in 2-7% cases.[4] Hepato splenomegally and elevated transaminases are relatively common.[4] Laboratory reports include leukopenia and elevated acute phase reactants. Anti-nuclear antibody is positive in about 7% of patients.[4] Histopathology with immunohistochemistry is the cornerstone in diagnosis, which shows paracortical coagulative necrosis distorting the nodal architecture; large number histiocytes admixed with CD8 positive T lymphocytes and paucity of neutrophil and plasma cells.[4] Granulomas are characteristically absent in Kikuchi's disease differentiating it from tuberculosis.[4] SLE lymphadenitis demonstrates degenerated nuclear material in walls of blood vessels, abundant plasma cells and pericapsular inflammation. The large cells observed in Kikuchi's disease are positive for CD68, MPO, and Ki-M1P1, while they are negative for CD3, CD15, CD20, CD30 or mucin, which excludes the possibility of lymphomas.

The disease is self-limiting and treatment is supportive with nonsteroidal anti-inflammatory drugs. Corticosteroids, hydroxychloroquine, methotrexate, and intravenous immunoglobulin have been used for severe cases.[5] It usually resolves in several weeks to months with a 1-year recurrence rate of 3-4%. We report this case for the clinicians to prevent misdiagnosis and inappropriate treatment of Kikuchi's disease especially in tubercular endemic areas and for some of its atypical features.

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