Abstract

Background: Kikuchi–Fujimoto disease, which was originally described in young women, is a benign condition characterized by necrotizing lymphadenitis and fever. Even though the clinical course is usually self-limiting, it can be associated with recurrences and rarely can be associated with systemic lupus erythematosus or can be complicated with hemophagocytic lymphohistiocytosis. We report the case of a 17-year-old Sri Lankan Sinhalese schoolboy who presented with fever and cervical lymphadenopathy diagnosed as Kikuchi–Fujimoto disease and was complicated with hemophagocytic lymphohistiocytosis subsequently. Later he fulfilled the criteria for systemic lupus erythematosus.

Case presentation: A 17-year-old previously healthy Sinhalese schoolboy presented with high-grade fever associated with chills and rigors associated with loss of appetite and loss of weight for more than 40 days. On examination, he had bilateral firm matted tender cervical lymphadenopathy and firm hepatomegaly. An excision biopsy of his right cervical lymph node revealed necrotizing lymphadenitis and immunohistochemistry of a lymph node biopsy favored Kikuchi disease. Initial antinuclear antibody and anti-double-stranded deoxyribonucleic acid tests were negative and his C3 and C4 levels were normal. An infections screening was negative. He was treated with steroids. While in hospital he developed hemophagocytic lymphohistiocytosis and renal impairment. Later his antinuclear antibody titer became positive in 1:160 and fulfilled the diagnostic criteria for systemic lupus erythematosus. He was managed with steroids and immune suppressive drugs and showed remarkable improvement.

Conclusion: Although Kikuchi–Fujimoto disease is uncommon in male patients, it needs to be considered in patients with lymphadenopathy and fever. The disease can be complicated with hemophagocytic lymphohistiocytosis and the patients need continuous monitoring for the possible development of systemic lupus erythematosus later in the course.

Keywords: Kikuchi–Fujimoto disease, Systemic lupus erythematosus, Hemophagocytic lymphohistiocytosis