

Abstract

Adult onset still's disease is a rare connective tissue disorder with a poorly understood aetiology and it is a leading cause for acquired macrophage activation syndrome which is a potentially life-threatening condition. In this case report we describe a 25 year old patient who presented with prolonged fever with arthralgia, myalgia and sore throat. His clinical examination revealed mild cervical lymphadenopathy and mild hepatosplenomegaly. He had neutrophil leucocytosis with very high Erythrocyte sedimentation rate, C reactive protein and serum ferritin with negative septic screening, autoimmune screening and haematological malignancy. After thorough investigation the diagnosis of adult onset still's disease was made according to Yamaguchi criteria. He did not respond to NSAIDS. He poorly responded to 3 days of intravenous steroids and fever recurred. Disease process was complicated with macrophage activation syndrome and treated with a prolonged course of intravenous methylprednisolone. He was responsive for high dose oral steroids, but he was a difficult case for steroid sparing with methotrexate. He ultimately was a candidate for biologics and needed tocilizumab (a recombinant, humanized, anti-human interleukin 6 (IL-6) receptor monoclonal antibody) injection for disease control.