Introduction

Adult-onset Stills disease (AOSD) is an autoimmune inflammatory disorder of unknown aetiology, characterized by quotidian fever (fever occurring daily), arthritis and an evanescent rash [1]. The clinical course of AOSD can be categorized into two main patterns: systemic pattern and chronic articular pattern. In patients with the systemic pattern, the disease course typically lasts only weeks to months and systemic features including fever, rash, serositis and hepatosplenomegaly are predominant. Patients with the chronic articular pattern have persistently active disease in which articular symptoms predominate, and this usually leads to destructive arthritis [2,3,4].

The major clinical features of AOSD are fever, arthritis and skin rash, which occur in about 75-90 percent of patients. Minor clinical features of AOSD include hepatomegaly, pleuritis, myocarditis and pericarditis [5]. Parenchymal lung involvement in AOSD is rare and encompasses a wide spectrum of disease including idiopathic interstitial pneumonia, nonspecific interstitial pneumonia, pleuritis, pleural effusion, atelectasis, transient pulmonary infiltrates and acute respiratory distress syndrome (ARDS) [6]. Macrophage activation syndrome (MAS) is a rare, potentially life-threatening complication of AOSD [5].

Laboratory findings in AOSD are nonspecific. A rise in acute phase reactants such as ESR, CRP, and serum ferritin are observed along with a neutrophil predominant leukocytosis with some immature cells. An elevated serum ferritin can be a striking feature and is commonly seen at levels above those typical of an acute phase response to other disorders [5].