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Case Report

Fuo Dilemma - Still Considering Still's Disease?

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Introduction

First described in children by George Still in 1896, "Still's disease" is eponymous with systemic Juvenile Idiopathic Arthritis (JIA) in persons >16 years of age. The term "adult Still's disease" has been used to describe a condition in adult patients who have features similar to children with JIA (usually fevers, arthralgias and rashes) who do not fulfill criteria for classic rheumatoid arthritis [1]. The etiology of Adult Still's Disease (ASD) is unknown; both underlying genetic factors (especially HLA-B17, -B18, -B35 and -DR2) [2] and infectious triggers (including *Yersinia enterocolitica* and *Mycoplasma pneumoniae*) [3] have been suggested. ASD is very rare with a prevalence estimated at 0.16 cases per 100,000 people [4]. It has a bimodal age distribution with peaks between 15 and 25 years of age and then between 36 and 46, with roughly equal distribution between both sexes.

Keywords: Adult Onset Still's Disease; Fever; Infection; Pharyngitis; Rash

Case Description

Our patient is a 23 years old previously healthy male who recently moved from Saudi Arabia to the US, who presented with complaints of intermittent objective fevers over the last three months associated with migratory pink rashes on his back and extremities that would develop acutely and completely resolve when the fever would subside. He also complained of migratory arthralgias in his wrists, knees, and small hand joints. Because he had intermittent nausea and vomiting with loose stools, he had been repeatedly diagnosed with viral gastroenteritis during prior multiple ED visits and treated symptomatically. On admission, he was febrile (38.1), tachycardic (132 bpm), and borderline hypotensive (107/59). Physical exam was remarkable only for mild pharyngeal congestion and a faint pink papular rash on his anterior chest and legs. Lab work was significant for neutrophilic leucocytosis (WBC of 26.9), transaminitis (AST 218, ALT 310), mild conjugated hyperbilirubinemia (T. Bili 1.6), and an elevated CRP (4.5). Workup for possible infectious agents was -ve. Autoimmune workup was remarkable for a weakly positive RF, a negative Anti-CCP, and a negative ANA. The serum Ferritin level was strikingly elevated at 36,388, which is 94 times the upper limit of normal. The patient was diagnosed with Adult onset still's disease and was started on prednisone 0.5 mg/kg/day with marked improvement in his symptoms within a few days.

Discussion

Since 1992, the diagnosis of ASD has been made using the Yamaguchi criteria [5]. Making a diagnosis of ASD requires the presence of five features, with at least two being major diagnostic criteria Table 1.

Major criteria	Minor Criteria
•Fever of at least 39°C (102.2°F) lasting at least one week •Arthralgias or arthritis lasting two weeks or longer •A nonpruritic macular or maculopapular skin rash that is salmon-colored in appearance and usually found over the trunk or extremities during febrile episodes •Leukocytosis (10,000/microL or greater), with at least 80 percent granulocytes	•Sore throat •Lymphadenopathy •Hepatomegaly or splenomegaly •Abnormal liver function studies, particularly elevations in aspartate and alanine aminotransferase and lactate dehydrogenase concentrations •Negative tests for Antinuclear Antibody (ANA) and Rheumatoid Factor (RF)

Table 1: Major and minor criteria.

Our patient had 4 major and 2 minor criteria, fulfilling the diagnosis of adult onset Still's disease. Although the major clinical features of Adult Still's Disease (ASD) are fever, rash, and arthritis or arthralgia (each occur in about 75 to 95 percent of patients), a full spectrum of Adult's onset Still's still can present with a large array of symptoms, signs and laboratory findings including:

-Myalgias: often worse with fever spikes, and can sometimes be severe and debilitating.

-Acute nonsuppurative pharyngitis: can occasionally precede the development of fever or rash and can also occur with disease relapses, estimated to occur in 69% of cases [6].

-Hepatomegaly: reported in 12-45% of patient according to multiple different studies [7].

-Cardiopulmonary disease: Pericarditis, pleural effusions, and transient pulmonary infiltrates have been observed in 30 to 40 percent of patients [8,9].

-Tender Lymphadenopathy and splenomegaly: enlarged cervical lymph nodes are seen in about 50% of patients. Splenomegaly may also occur in one-third to one-half of patients with ASD.

-Elevated serum ferritin: ASD has been associated with markedly elevated serum ferritin concentrations in as much as 70 percent of patients. This is probably an acute phase response, since hepatocytes responding to inflammatory cytokines can increase ferritin synthesis [10]. The elevations correlate with disease activity and have been suggested as a serologic marker to monitor the response to treatment [11-13].

Rare as it is, ASD should be considered when other more common causes of Fever of unknown origin are not consistent with the patient's picture, especially if they are young and/or have a history of long-term or intermittent symptoms.

Funds: N/A

Conflict of interest: None

Due to the absence of any patient's identifiers in the case description, neither an ethics approval nor a patient's consent was deemed to be necessary, in accordance to our institution's (Riverside Community Hospital) policy.

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