

Just Another Fever and Rash? A Broad Differential Leading to a Relatively Uncommon Condition in Adult Still's Disease.

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Just Another Fever and Rash? A Broad Differential Leading to a Relatively Uncommon Condition in Adult Still's Disease

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INTRODUCTION

Rashes are a common complaint in the inpatient and outpatient setting. Adult Still's Disease (ASD) is a relatively uncommon diagnosis that often presents with a rash. There have been few published reports of the prevalence of ASD, however, one group reported a crude prevalence of 1.5 cases per 100,000–1,000,000.¹ Patients usually present with a group of signs and symptoms that will point toward the diagnosis.

CASE REPORT

A 38-year-old female with no significant past medical history presented to the hospital after having four weeks of bilateral ankle and foot pain that progressed to include all joints. She was having nightly fevers with a t-max of 105. She also reported an intermittent, mildly pruritic rash (Image 1) mainly under her breasts that she attributed to a yeast infection or heat rash secondary to the fevers. The rash began to spread over her extremities and palms. She had tried a prednisone taper, meloxicam, and doxycycline for presumed Lyme disease without any improvement in her symptoms. She was being worked up as an outpatient but began having pleuritic chest pain, which brought her into the hospital. A CTA scan (Image 2) of the chest was performed which revealed several prominent mediastinal and axillary lymph nodes along with a moderate pericardial effusion. An echocardiogram (Image 3) was performed confirming the pericardial effusion without tamponade physiology.

Labs included:

- ▶ WBC 14.3 (82% granulocytes)
- ▶ Ferritin 576
- ▶ CRP 328 and ESR 122
- ▶ AST 106 ALT 148
- ▶ ACE, ANA, anti-CCP, RF, HLA-B27, EBV, Hepatitis panel, G6PD and SPEP were all negative.

Using the Yamaguchi criteria (Table 1), the patient was diagnosed with ASD. She subsequently was placed on prednisone 1mg/kg/day and began to have resolution of her symptoms. She was discharged from the hospital a couple of days later with rheumatology follow-up.

YAMAGUCHI CRITERIA³

Major Criteria	Minor Criteria
Quotidian fevers of at least 39° Celsius (102.2° Fahrenheit)	Sore throat
At least two weeks of arthralgias	Lymphadenopathy
Salmon-colored rash generally on the trunk or extremities during fevers	Hepatomegaly or splenomegaly
Leukocytosis with at least 80% granulocytes	Abnormal liver function tests
	Negative ANA and RF

Requires 5 features to be positive, 2 of which need to be major criteria.

Table 1



Image 1. Salmon-colored rash² commonly seen in Adult Still's Disease demonstrating what would be found on the patient in the case.

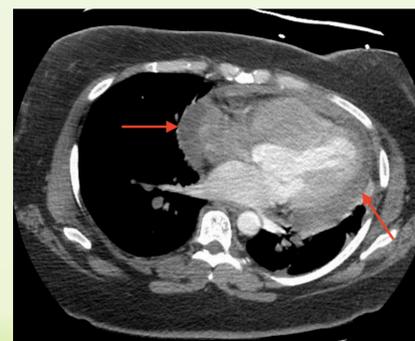


Image 2. Pericardial effusion seen on CT imaging

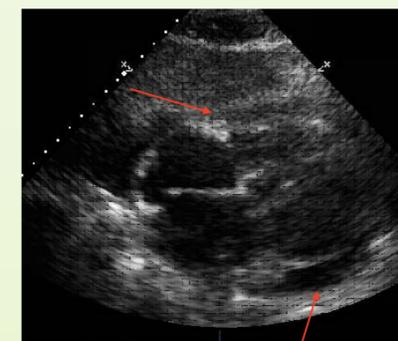


Image 3. Pericardial effusion seen on echocardiogram

DISCUSSION

- ▶ Although ASD is relatively uncommon, it is important to be able to recognize the disease in order to give the patient the best and most effective treatment possible as early intervention can potentially improve prognosis.⁴
- ▶ Treatment depends on the severity of the disease:⁵
 - Mild disease presents with fevers, mild rash, and mild arthritis. Treat with NSAIDs (duration 4 weeks–3 months) with response rates in 20% of cases. Add low dose steroids if not responsive.
 - Moderate disease presents with daily high fevers, debilitating joint pain, non-threatening internal organ involvement. Treat with prednisone 0.5-1 mg/kg (duration until symptoms resolve with a 2–3 month prolonged taper).
 - Severe disease includes cardiac tamponade, severe hepatic involvement, and DIC. Treat with high dose IV pulse steroids followed by high dose oral steroids and anakinra (few case reports support choice of anakinra)

- ▶ Although ferritin, CRP, and ESR are not a part of the Yamaguchi criteria, they are often elevated in ASD as they are acute phase reactants. Ferritin may be considered as a marker for monitoring response to treatment.⁶

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