CASE REPORT

Stay Still’s for POCUS: using bedside ultrasound to screen for cardiac complications

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Abstract

Adult Still’s disease (ASD) is a rare systemic inflammatory disorder of unknown etiology most commonly characterized by daily spiking fevers, an evanescent, ‘salmon-colored’ rash, and arthralgia. Cardiac complications such as pericarditis, myocarditis, heart failure, and pericardial effusion progressing to tamponade have been reported. Because of the severe and potentially lethal complications associated with these processes, the clinician’s index of suspicion must remain high and the threshold for cardiac imaging low. Here, we present a case of ASD-associated myocarditis identified quickly by point-of-care ultrasound, allowing for prompt workup and treatment.

INTRODUCTION

Adult Still’s disease (ASD) is a rare systemic inflammatory disorder characterized by daily spiking fevers, an evanescent, ‘salmon-colored’ rash and arthralgia. Other frequently involved organs include the lungs, liver and lymphoid tissues [1]. Although rare, significant cardiac involvement leading to pericarditis, myocarditis and pericardial effusion progressing to tamponade have been reported [2]. Because of the severe and potentially lethal complications associated with these processes, the clinician’s index of suspicion must remain high and the threshold for cardiac imaging low. Here, we present a case of ASD-associated myocarditis identified quickly by point-of-care ultrasound (POCUS), allowing for prompt workup and treatment.

CASE REPORT

A previously healthy 27-year-old Hispanic male presented to the emergency department (ED) reporting abrupt onset quotidian fevers as high as 39.1°C and an evanescent rash that started one week prior. He noticed that the rash coincided with fevers and worsened with hot showers. He subsequently developed lower extremity weakness and myalgias on day three of fevers, followed by headache, odynophagia, vomiting, cough and dyspnea on day four. On day seven, he experienced pleuritic chest pain and palpitations, which prompted his ED visit. Of note, the patient was in a monogamous relationship and worked as a counselor for elementary school children. He had no prior medical problems and apart from a daily multivitamin was not taking any medications. He
Table 1: Laboratory values from the initial workup and their corresponding reference ranges

| Laboratory study           | Value          | Reference range       |
|----------------------------|----------------|-----------------------|
| White blood count          | 19 500/μL†     | 4500–10 500/μL        |
| Absolute neutrophil count  | 17 500/μL†     | 1600–7300/μL          |
| High sensitivity troponin-I| 679 pg/mL†     | <18 pg/mL             |
| Ferritin                   | 8390 ng/mL†    | 30–300 ng/mL          |
| C-reactive protein         | 289.7 mg/L†    | <5 mg/L               |
| Erythrocyte sedimentation rate | 60 mm/hr† | 0–20 mm/hr            |
| AST                        | 55 IU/L†       | 5–40 IU/L             |
| B-natriuretic peptide      | 1039 pg/mL†    | 5–100 pg/mL           |
| CSF white cell count       | 1/mm³          | 0–10/mm³              |
| CSF glucose                | 79 mg/dL       | 40–70 mg/dL           |
| CSF protein                | 25 mg/dL       | 15–45 mg/dL           |

Figure 1: The patient presented with a diffuse, confluent, erythematous, maculopapular rash involving the palms, which waxed and waned with fevers.

specifically had no personal or family history of inflammatory arthritis.

In the ED, the patient was febrile to 39°C and tachycardic with a heart rate of 150 bpm, a blood pressure of 140/90, and oxygen saturations >95%. On examination, a diffuse, confluent maculopapular rash was noted (Fig. 1). Range of motion was limited by severe proximal myalgias. Synovitis of the metacarpophalangeal and proximal interphalangeal joints of the hands was appreciated. Cardiac examination revealed tachycardia with easily auscultated heart sounds and the absence of a friction rub. Assessment of jugular venous pressure was limited by the patient’s body habitus. A troponin was drawn due to concern for myocardial infarction and pulmonary embolism, and a lumbar puncture was performed due to high fever, leukocytosis and headache. Pertinent laboratory values are presented in Table 1.

An EKG showed sinus tachycardia at a rate of 145 BPM without ST-changes or T-wave inversions (Fig. 2).

Due to the patient’s pleuritic pain and tachycardia, a CT angiogram of the chest was obtained which showed no pulmonary embolism but revealed trace pericardial effusion with minimal pericardial thickening as well as lower lobe ground-glass opacities with bronchial thickening indicating inflammation. Enlarged axillary lymph nodes were noted bilaterally with cortical thicknesses up to 2.1 centimeters.

Given the constellation of historical, clinical and laboratory findings, an infectious versus rheumatologic etiology with cardiac involvement was suspected. A cardiac POCUS exam was performed to look for evidence of effusion, tamponade, myocarditis or heart failure. A Sonosite X-Porte 5-1 MHz phased-array transducer was used. Parasternal long-axis view revealed a trace anterior pericardial effusion measuring 4.6 mm. There was no swinging of the heart within the pericardial sac and no systolic atrial collapse or diastolic right ventricular collapse suggestive of tamponade physiology. There was diminished left ventricular squeeze with tachycardia, and no wall motion abnormalities were seen. On short-axis view, a thickened inteventricular septum (IVS) and left ventricular posterior wall (LVPW) were noted, measuring 15.1 and 16 mm, respectively, at end-systole. The normal IVS and LVPW thickness in this age group (20-29) was 8.3 and 7.5 mm, respectively [3]. There was no right ventricular dilation, but a pseudo-D-sign was present due to marked septal thickening (Fig. 3).

Our bedside POCUS in context with the patient’s clinical picture led to prompt rheumatology consultation and more timely testing to exclude malignancy and infection. Shortly afterwards, a lymph node biopsy was negative, ruling out lymphoid malignancy. Serologic testing for an array of infectious diseases was also unrevealing, allowing for the diagnosis of ASD by the Yamaguchi Criteria. He was subsequently treated with prednisone 0.5 mg/kg daily with rapid improvement in symptoms and cardiac function. Contrast-enhanced transthoracic echocardiogram (TTE) after 2 weeks of treatment showed normalization of the myocardium with an ejection fraction of 60–65% and resolution of the septal flattening initially seen on bedside POCUS (Fig. 4). The IVS and LVPW measure 10 and 11 mm, respectively.

DISCUSSION

Bedside POCUS is a reliable diagnostic tool that can be performed and interpreted by the internist within minutes to assess the heart in real time [4]. In comparison, formal TTE and cardiac magnetic imaging are more time-consuming, costly, and resource-intensive imaging modalities [5, 6]. We encourage the
Figure 2: An EKG was obtained, which showed a long RP (likely sinus) tachycardia at a rate of 145 BPM with subtle ST-segment elevations in leads V2 and V3 not meeting criteria for ACS in a male <40 and without T-wave inversions.

Figure 3: Parasternal short-axis view of the heart obtained with a 5–1 MHz phased-array transducer at the level of the papillary muscles shows a thickened interventricular septal wall measuring 15.1 mm as well as a thickened left ventricular posterior wall measuring approximately 16 mm.

routine use of bedside ultrasound in patients with suspected systemic inflammatory disease as an initial step to evaluate for potentially life-threatening cardiac manifestations, as it can cost-effectively rule out cardiac abnormalities while also identifying patients in need of more detailed and expensive imaging.

In our case, bedside ultrasound revealed a strikingly edematous myocardium. This wall thickening without chamber dilatation is characteristic of fulminant myocarditis, in which patients rapidly develop heart failure, but afterwards have nearly complete return of cardiac function, as was the case here. A study investigating myocardial thickness in fulminant myocarditis showed that on days 1–3 after onset, the average IVS and LVPW measured 14.6 mm (SD 3.7 mm) and 13.0 mm (SD 2.9 mm), respectively [7]. While this has been described for idiopathic myocarditis, it has not been characterized on POCUS in ASD-associated myocarditis. Our patient’s IVS and LVPW measured 15.1 and 16 mm.

Patients in whom a systemic inflammatory disease is suspected with symptoms, labs and examination findings concerning for cardiac involvement can benefit from prompt imaging of the heart with bedside ultrasound.
Figure 4: Contrast-enhanced TTE 2 weeks after initiation of prednisone shows resolution of the septal flattening seen on initial bedside imaging as well as ongoing resolution of myocardial thickening. The interventricular septal wall measures 10 mm, and the left ventricular posterior wall measures 11 mm.

CONFLICT OF INTEREST STATEMENT
The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

FUNDING
No funding or financial support was received for the conceptualization, writing or submission of this project.

ETHICAL APPROVAL
The research presented in this paper was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Ethics approval was not required for the research presented in this paper. All images and descriptions have been deidentified.

CONSENT
All images and descriptions have been deidentified; the right of privacy has not been infringed upon in this manuscript or in the preparation of this manuscript. The authors confirm that informed consent was obtained from the patient. Measures have been taken to protect the patient's anonymity including omission of the patient's name, initials, medical record number and other identifiable characteristics from the manuscript and figures. An informed consent document is on file with the authors, which conforms with HIPAA compliance standards. The informed consent was documented using the Oxford University Press Patient Consent Form.

GUARANTOR
The authors nominated Dr. Christopher T. Kelly as the guarantor of this manuscript.

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