A case of myopericarditis and catastrophic antiphospholipid syndrome (CAPS)

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Background: Myopericarditis secondary to catastrophic antiphospholipid syndrome (CAPS) is an extremely rare and life-threatening condition. . Clinical suspicion and early diagnosis is vastly important to ensuring proper therapy and management. We present a case of CAPS and myopericarditis.

Case: A 21-year-old male presented with 1 week of worsening shortness of breath, orthopnea and bilateral leg swelling. In the ER he was hypoxic to the 80s and had elevated diastolic pressures to the 120s. Chest x-ray showed bilateral pulmonary infiltrates compatible with pneumonia or edema. EKG showed sinus tachycardia without ST deviations. Bedside echocardiogram showed EF of 15%. Labs were notable for elevated troponin of 8.9ng/mL and BNP of 89,000pg/mL. His past medical history was notable for Systemic Lupus Erythematosus, Antiphospholipid syndrome and cleft mitral valve repaired with annuloplasty ring. He also had recent history of testicular necrosis, small bowel ischemia and other ischemic events supporting his antiphospholipid diagnosis. The patient was being treated with hydroxychloroquine, mycophenolate mofetil, prednisone and warfarin as an outpatient. He underwent right and left heart catheterization showing normal coronaries and WHO classification group 2 pulmonary hypertension with elevated pulmonary artery pressures (systolic PA: 62mmHg; mean PA: 46mmHg) elevated wedge 31mmHg and LVEDP 24mmHg. Further lab testing revealed positive dsDNA, low complement and positive antiphospholipid Abs. Cardiac MRI was completed which showed delayed enhancement pattern consistent with myopericarditis. RV biopsy was done later when the patient was stabilized and pathology was sent but showed no evidence of myocarditis. The patient was diagnosed with Lupus myocarditis and catastrophic antiphospholipid syndrome (CAPS). Patient was treated with steroids, cyclophosphamide, plasma exchange and afterload reduction and therapeutic anticoagulation with unfractionated heparin. He had a prolonged ICU stay including VV ECMO, multiple intubations and septic shock. . The patient's ejection fraction improved over his hospital course and peaked at 30-35%. He was discharged in stable condition.

Decision-Making:
The patient's presentation of acute coronary syndrome and acute systolic heart failure prompted him to undergo coronary evaluation and right heart catheterization with endomyocardial biopsy. Given his history of active lupus,

antiphospholipid syndrome and recent ischemic events, further rheumatologic evaluation was indicated along with cardiac MRI to help determine an etiology. Less than 1% of patients with APS develop CAPS and even less will present with acute myopericarditis and acute systolic heart failure leading to cardiogenic shock. Classic CAPS is characterized with rapid, diffuse vascular thrombosis and ischemia. This mechanism of small vessel thrombosis and ischemia was thought to be the mechanism for myopericarditis, elevated troponins, acute systolic heart failure and fulminant cardiogenic shock in our patient. Prompt diagnosis was crucial in this case as it directed medical therapy. In these cases an endomyocardial biopsy is a Class IIA indication, although the diagnostic yield is only about 10- 30%. Due to his instability however, the patient was unable to undergo this procedure until after being treated empirically for the presumed diagnosis.

Conclusions:
CAPS should be suspected in patients with H/O APS presenting with acute myopericarditis and/or acute systolic heart failure. The diagnosis and treatment of such cases is complex but the outcome does not need to be always fatal. We feel this case report will help inform the community of this deleterious disease process and assist with diagnosis and management of future cases.

APPENDIX:
Images 1 and 2:
Short axis and 2 chamber myocardial delayed enhancement images showing transmural and subepicardial delayed enhancement in the basal/mid inferior wall

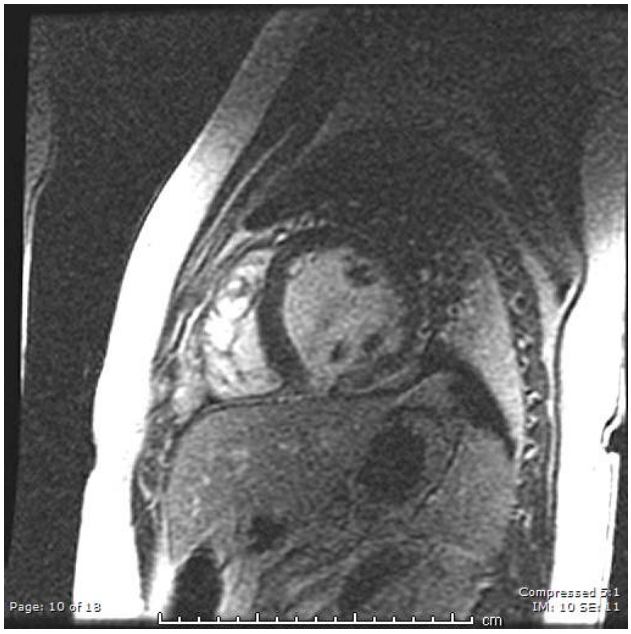

Image 1

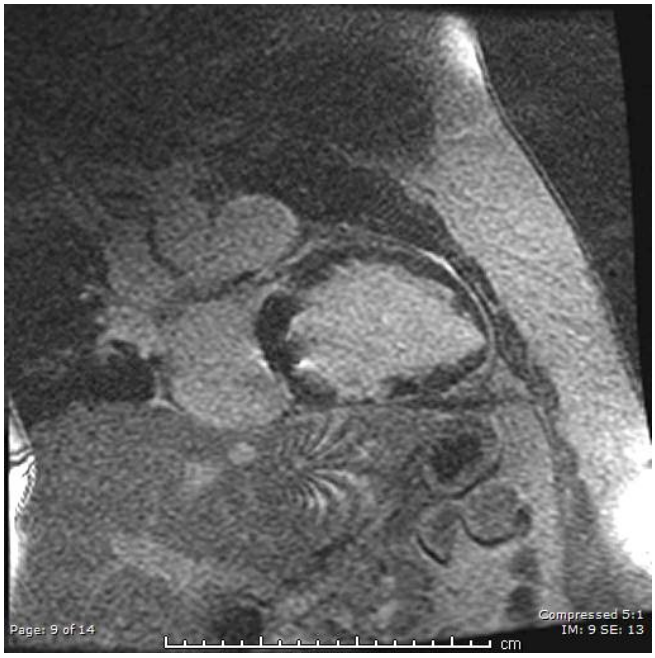

Image 2

Images 3 and 4: Short axis saturation recovery single-shot acquisition (SASHA) T1 map showing elevated myocardial T1 of 1299ms (normal 1050ms).

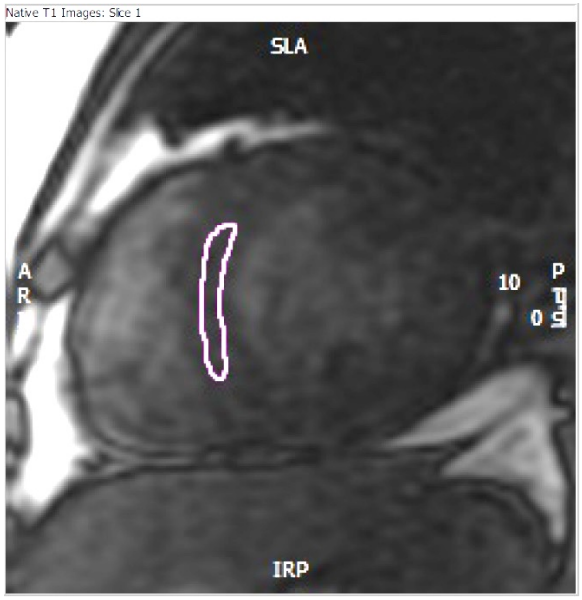

Image 3

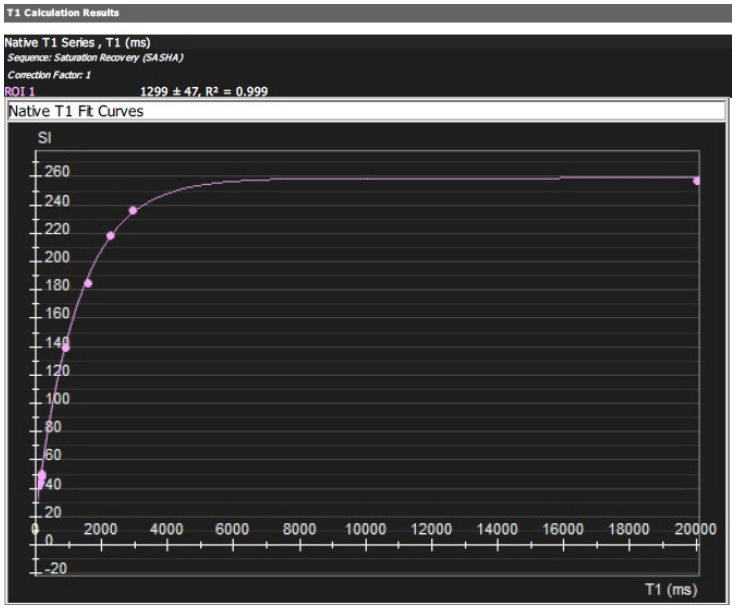

Image 5: Endomyocardial biopsy showing normal cellular architecture and no evidence of infiltration, inflammation, necrosis or disarray.