Shocked to Discover Lupus: Rapid Progression of Effusions and Renal Failure in Undiagnosed Systemic Lupus Erythematosus

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Background
- Systemic Lupus Erythematosus (SLE) is a rheumatologic disease that can affect multiple organ systems with a wide variation in manifestations and time course of symptoms
- Pleural involvement and pericarditis with or without effusion occur in about 93% and 25% of patients, respectively
- Renal involvement is present in about 50% of cases
- The above manifestations typically take several months to cause significant morbidity or mortality

Case Overview
- 22-year-old female patient with no known SLE history who presented initially with mild acute kidney injury (AKI) and small pleural/pericardial effusions
- SLE workup was sent on initial hospitalization with positive results but the patient did not follow up as an outpatient with rheumatology
- She was subsequently hospitalized only 6 weeks later with life-threatening effusions and hyperkalemia with resulting cardiac arrest
- Patient also suffered renal failure requiring hemodialysis

Initial Presentation
- 22-year-old female admitted for evaluation of hypertension and hypothyroidism as well as AKI and metabolic acidosis, also positive for Covid-19
- Small, non-concerning pericardial and pleural effusions identified (see images)
- Serum creatinine was 1.46 mg/dL with eGFR of 52 mL/min/1.73m² on presentation
- Creatinine was normal with eGFR at 82 at discharge
- Initial SLE Antibody workup:
  - Positive ANA with 1:640 titer and speckled pattern
  - Positive anti DNA Ab multiplex
  - C₃ 21 mg/dL (normal range 87-200)
  - C₄ < 8 mg/dL (normal range 19-52)
  - Indeterminate p-ANCA

Repeat Admission
- About 6 weeks later she presented to the ED with SOB associated with cough, fever, and diarrhea; she was in respiratory failure with active vomiting and suspected aspiration after which she lost consciousness and was found to be pulseless
- Ventricular fibrillation was identified and shock administered; patient achieved ROSC with CPR
- Post-arrest EKG did not suggest ischemia
- Creatinine at this time was 1.72 with eGFR at 43; potassium was 8.6
- CT of the chest showed large bilateral pleural effusions with complete collapse of the lung on the left, as well as large pericardial effusion and small abdominal ascites
- Patient spent 2 days in the MICU during which her effusions were drained and hemodialysis was initiated
- Renal biopsy revealed diffuse proliferative glomerulonephritis with membranoproliferative features consistent with lupus nephritis class IV
- Further immunologic workup:
  - RNP IgG >8.0
  - Ro IgG 4.3
  - Scl IgG 1.3, Sm IgG at >8.0

Resolution
- On further questioning, patient reported history of malar rash, oral ulcer, and alopecia
- She was initially treated with IV solu-medrol and discharged on high-dose oral prednisone as well as mycophenolate mofetil and hydroxychloroquine with instructions to follow up with rheumatology
- On discharge, eGFR was 65 with potassium at 4.1 Patient was discharged without further need for dialysis

Discussion
- While significant pleural/pericardial effusions and fulminant renal failure can develop in patients with SLE, it is rare for all three to develop rapidly over the course of weeks
- This patient was found to have relatively mild AKI with small, non-concerning pericardial as well as unilateral pleural effusions on initial presentation with positive ANA, low complement levels and anti DNA Ab
- Only six weeks later she was diagnosed with massive bilateral pleural effusions and large pericardial effusion, as well as renal failure requiring hemodialysis
- Patient also suffered cardiac arrest secondary to hypoxia vs. hyperkalemia vs. cardiac tamponade
- SLE is a highly variable disease with a multitude of presentations and no standard time course
- This case, while it represents an unusual disease course, demonstrates the importance of rapid identification and treatment of complications of the disease with close follow-up following positive results on initial workup

Images
- Initial Presentation - CTA chest PE protocol
- Repeat Admission - CTA chest

Works Cited