Sarcoidosis presenting with nephrotic syndrome

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ABSTRACT:

We describe 39-year-old Caucasian man presenting with pulmonary embolism, inferior vena cava thrombosis, nephrotic syndrome and mediastinal lymphnodes. Mediastinal lymphnodes biopsy through mediastinoscopy revealed numerous non-necrotizing granulomata establishing the diagnosis of sarcoidosis. There are few cases reported of sarcoidosis presenting with such extensive emboli due to nephrotic syndrome.

INTRODUCTION:

A case of sarcoidosis disease presenting with nephrotic syndrome and pulmonary embolism.

CASE DESCRIPTION:

A 39 year old caucasian male with no significant past medical history presented to emergency department complaining of right sided chest pain and fever for 3 days. He described his pain as sharp, radiating to the back and worsening with movement and breathing. Physical exam revealed normal appearing male with mild crackles on the right pulmonary field. Chest X-ray showed right lower airspace opacity and right pleural disease. Labs were consistent with increased D-dimer (3.71mg/L), hypoalbuminemia (1.6U/L), hypertriglyceridemia (419mg/dL), hypercholesterolemia (280mg/dL), normal AST/ALT (12/12U/L), normal BUN/creatinine (10/1.0mg/dL), proteinuria (>600), and 24-hour urine protein (>16gr). Nephrotic syndrome workup inclusive of HIV, hepatitis profile, autoimmune workup, renal ultrasound was negative. Workup for Goodpasture's syndrome was also negative. CTA-chest showed bilateral Pulmonary Emboli (fig 1) with pulmonary infarctions, large Inferior Vena Cava thrombus extending proximally into portal/hepatic system, mediastinal and hilar lymphadenopathy. The working diagnosis was Pulmonary Embolism secondary to Nephrotic Syndrome. He was started on anticoagulation, lasix, ACEI, statins. Bone marrow biopsy and flow cytometry to rule out any lymphoproliferative disorders resulted negative. Mediastinoscopy was performed and mediastinal lymphnodes biopsy revealed numerous non-necrotizing granulomata. The clinical and pathological findings were felt to be most consistent with an underlying diagnosis of sarcoidosis.

DISCUSSION:
Sarcoidosis is a multisystem granulomatous disorder of unknown etiology that most commonly affects young adults and is characterized pathologically by the presence of noncaseating granulomas in involved organs. Renal involvement in Sarcoidosis most commonly presents with granulomatous interstitial nephritis and calcium metabolism abnormalities such as hypercalcaemia, hypercalciuria. Glomerular involvement and nephrotic syndrome although rare may complicate Sarcoidosis.\(^1\) The pathogenesis of glomerular disease in sarcoidosis is not known nor has a causal relationship been proven. The risk of venous thrombosis and pulmonary emboli in nephrotic syndrome is greatest when there is heavy proteinuria and the serum albumin is less than 2. The presence of heavy proteinuria helps differentiating glomerulopathies from interstitial nephritis. Patients with nephrotic syndrome are at increased risk for venous thrombosis, particularly deep vein and renal vein thrombosis (DVT and RVT), which may result from loss of endogenous anticoagulant proteins such as AT III (antithrombin). Pulmonary embolism (PE) has been described in nephrotic patients with or without an evident DVT or RVT. The estimated prevalence of asymptomatic PE in patients with the nephrotic syndrome ranges from 12 to over 30 percent.\(^2\) The case is an unusual example of sarcoidosis and severe nephrotic syndrome complicated by a hypercoagulable state.

REFERENCES:
