An 18-year-old with nephrotic syndrome and chest pain: Questions

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Case presentation

A 15-year-old African American male presented with periorbital and lower extremity edema and was diagnosed with nephrotic syndrome (NS). Hepatitis B and C serologies, cytomegalovirus PCR and rapid human immunodeficiency virus tests were negative, and genetic studies revealed no abnormal variants in the NPHS2, ACTN4 or TRPC6 genes. The initial biopsy showed minimal change disease, although focal segmental glomerulosclerosis (FSGS) was suspected based on the patient's age at presentation. Subsequent biopsy confirmed the diagnosis of FSGS. He was treated with high-dose prednisone for 6 weeks, and his NS was steroid-resistant. He subsequently received an 8-week course of oral cyclophosphamide, which was prematurely stopped due to an increased urine protein:creatinine

ratio and worsening anasarca. He then received tacrolimus, which induced remission after 5 months.

Three years following his initial diagnosis of NS, he presented to our emergency room with a 1-week history of dry cough and 1 day of worsening severe substernal pleuritic chest pain. Further questioning revealed a medical history of 3 weeks of nightly fevers and sweats, 2 weeks of generalized abdominal pain, headaches and generalized fatigue. He denied any weight changes. He continued to take tacrolimus. Physical exam was unremarkable. Specifically, there was no edema, palpable lymphadenopathy or hepatosplenomegaly. He was afebrile, with a blood pressure of 94/57, heart rate of 112 beats per minute and respiratory rate of 19 breaths/minute. His urine protein:creatinine ratio was 0.1 mg/mg and serum albumin was 3.2 g/dL. His serum tacrolimus level was appropriate at 8.5 ng/mL and his serum creatinine was 0.8 mg/dL. He had no reaction on a tuberculin skin test. An electrocardiogram revealed sinus tachycardia.

The answers to these questions can be found at doi:[10.1007/s00467-013-2694-2](https://doi.org/10.1007/s00467-013-2694-2).

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Questions

1. What is the differential diagnosis and what studies would you perform next?
2. Are there any aspects of his history that predispose him to any particular diagnoses?
3. Is his history of nephrotic syndrome related to his new diagnosis?

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