Case 19

J.W. is a 48 year-old man who presented to CPMC with one day of hemoptysis (coughing up blood) and a 6 month history of headache, intermittent cough, arthralgias (joint pains), 10 pound weight loss and intermittent fevers. One month prior to admission during an evaluation of his headache he was found to have sinusitis that was treated with antibiotics without success. There was no other past medical history and he denied the use of tobacco, alcohol, or other drugs. There was no known exposure to tuberculosis. On physical exam he had râles in both lower lung fields, swelling of his knees and wrists, and an ulcer in the right nares. Pertinent laboratory studies showed a Hct 25% (normal > 36%), urinalysis with 3+ protein,¹ and 25-50 RBC per high powered field;² creatinine 3.7 mg/dl (elevated); normal liver function tests. His chest X-ray showed a diffuse infiltrate in the mid-lung fields bilaterally (Fig. 1) with several cavitory lesions confirmed by chest CT (Fig. 2). Skin tests were positive for Candida but a PPD (test for TB) was negative.

![Fig. 1. PA chest X-ray of J.W. on presentation to CPMC. Arrows point to possible cavitary lesions.](image)

Serological studies were negative for rheumatoid factor, ANA and anti-DNA antibodies. A c-ANCA was positive. Biopsy of the nasal ulcer and the lung yielded similar lesions showing vessel walls invaded by inflammatory cells including lymphocytes and monocytes. Some of the lesions appeared granulomatous; AFB (acid fast bacilli) and fungal stains were negative. The diagnosis of Wegener's Granulomatosis was made and the patient was treated with prednisone and cyclophosphamide. The patient improved and his renal function and chest X-ray returned to normal within several months of therapy.
Fig. 2. Chest CT of J.W. demonstrating cavitary lesion in left upper lobe of the lung (arrow).

1Indicative of renal disease characterized by abnormal loss of protein in the urine

2Hematuria, or blood in the urine, usually indicating intrinsic renal disease

Questions for Case 19

(1) How does the immunopathology of Wegener's granulomatosis differ from SLE or polyarteritis nodosa?

(2) What is known about the mechanism of granuloma formation? What is the differential diagnosis of granulomas in a pathological lesion?

(3) What is a c-ANCA and describe briefly how it is performed. What is a possible role of auto-antibodies in Wegener's?