Dyspnea and Hemoptysis Develop in a Young Man With Prostatitis*  

Carolina Q. See, MD; H. Ari Jaffe, MD; and Dean E. Schraufnagel, MD, FCCP

A 31-year-old man was admitted to the hospital with hemoptysis and progressive dyspnea. He was well until 4 months prior to hospital admission, when night sweats, anorexia, fevers, and 30-lb weight loss developed. Two months prior to hospital admission, he began to experience rectal pain, dysuria, and hematuria. One month prior to hospital admission, he presented with acute urinary retention at the emergency department. His prostate on digital rectal examination was enlarged and tender, and a CT of the pelvis suggested that he had prostatic abscess. A transurethral prostatic resection uncovered pockets of pus and friable tissues. Microscopically, acute and chronic necrotizing prostatitis was seen (Fig 1) He was discharged home on oral antibiotics.

Over the next 2 weeks, the patient had blurred vision, hearing loss, and myalgias. He returned to the emergency department and was administered ocular tobramycin. Findings of a chest radiograph done at that time were normal.

Four days prior to hospital admission, blood-tinged sputum and shortness of breath developed. This progressed, and the patient was admitted to the hospital.

The history of the patient revealed that he immigrated to the United States 12 years ago from Ecuador and had not traveled since then. He denied smoking, alcohol, or IV drug use. He also denied a history of tuberculosis or known exposure to tuberculosis. Results of a tuberculin skin test done 1 month before hospital admission were negative.

At the time of hospital admission, the patient was in mild respiratory distress and tachycardic. Examination of the chest was normal. Lateral episcleritis was noted in both eyes. His nasal turbinates were clear, and tympanic membranes were normal. Neurologic examination was grossly symmetrical and intact. Admitting laboratory data revealed hemoglobin of 8.2 g/dL, mean corpuscular volume of 78 femtoliters, a leukocyte count of 11,000/mL with 84% neutrophils, 8% lymphocytes, 5% monocytes, 2% eosinophils, and 1% basophils. The platelet count was 459,000/mL. With no supplemental oxygen, the arterial pH was 7.51, PaCO₂ was 29 mm Hg, and PaO₂ was 53 mm Hg. The BUN was 8 mg/dL, and creatinine was 0.7 mg/dL. The urinalysis showed “large” blood, “trace protein,” 40 RBCs per low-power field, and 18 WBCs per low-power field. A chest radiograph showed the development of bilateral upper lobe alveolar opacities that were not present 1 week before (Fig 2).

The patient was placed on respiratory isolation, and hypoxemic respiratory failure rapidly developed,
requiring mechanical ventilation. His repeat chest radiograph showed rapid and extensive progression to nearly complete alveolar opacification (“white-out”) with air bronchograms.

What is the diagnosis?
Diagnosis: Wegener granulomatosis with diffuse alveolar hemorrhage and prostatitis

First described in 1936, Wegener granulomatosis is a clinical syndrome that consists of necrotizing granulomatous vasculitis of the upper respiratory tract, lungs, and kidneys. The limited form of Wegener granulomatosis spares the kidney. Wegener originally described it as a rhinogenic form of polyarteritis nodosa due to its prominent nasal and paranasal involvement. Although the clinical attention has been focused mainly on the respiratory and renal systems, Wegener granulomatosis has been known to involve almost any organ in the body, including the eyes, ears, heart, skin, joints, peripheral and central nervous systems, and lower genitourinary tract. It can affect persons of all ages but is more common in middle-aged patients, predominantly men.

An immunologic pathogenesis has been implicated because of the findings of hypergammaglobulinemia and circulating antibodies, particularly anti-neutrophil cytoplasmic antibodies in patients with this disorder. The hypothesis is that the antibodies induce a necrotizing vasculitis by inciting a respiratory burst and degranulation of neutrophils and monocytes. These initial events require the priming of leukocytes by cytokines, leading to the expression of proteinase 3 and myeloperoxidase on the cell surface.

Lung involvement occurs in 80% of cases of Wegener granulomatosis. Its involvement may range from minimal to life threatening. Symptoms are nonspecific and include cough, dyspnea, hemoptysis, and pleuritic chest pain. Cough is usually nonproductive and is the most frequent symptom. Dyspnea occurs particularly with diffuse alveolar hemorrhage and also in the rare setting of tracheal involvement. Pulmonary capillaritis is seen in approximately one third of patients with lung disease and may lead to diffuse alveolar hemorrhage, hemoptysis, and rapidly changing alveolar opacities on chest radiograph.

Although the lung is the most common organ system affected in Wegener granulomatosis, diffuse alveolar hemorrhage as a result of capillaritis is uncommonly seen. Hemoptysis, if it occurs, is usually related to nodules, inflammatory, or necrotic lesions, rather than diffuse alveolar hemorrhage. In the largest surgical pathologic series of Wegener granulomatosis, alveolar hemorrhage was the dominant pathologic finding in only 7% of all 87 specimens. When diffuse alveolar hemorrhage is present, it is often the initial manifestation and is usually associated with renal involvement. It may rapidly progress to respiratory failure with a mortality of approximately 50%. On the chest radiograph, a diffuse, bilateral alveolar filling pattern is seen. Our patient did not initially present with diffuse alveolar hemorrhage and did not have renal impairment.

Diffuse alveolar hemorrhage is a cause of elevated carbon monoxide diffusing capacity. However, this test is usually not performed, as dyspneic patients in the intensive care setting poorly tolerate it. The findings of flexible fiberoptic bronchoscopy with BAL are distinctive in alveolar hemorrhage. The lavage fluid is bloody, and iron stains of bronchoalveolar specimens show hemosiderin-laden macrophages indicative of chronic hemorrhage.

Kidney involvement is common and usually occurs within 2 years of the initial upper airway disease. Involvement of the lower urogenital tract in Wegener granulomatosis is rare. It may manifest itself as acute urinary retention or prostatitis, orchitis, ureteral stenosis, bladder pseudotumor, and penile ulceration. Prostatitis is the most common manifestation of lower urogenital disease, causing urinary frequency, dysuria, gross hematuria, and urinary retention. However, it is usually asymptomatic; symptomatic prostatitis is reported to occur between 2.3% and 7.4% of patients with urogenital involvement. Urogenital symptoms usually resolve promptly with high-dose corticosteroids and immunosuppressive drugs such as cyclophosphamide.

In a review of 80 patients with Wegener granulomatosis, the main pulmonary features of nodular opacities usually were combined with urogenital symptoms. We could find no report on the combination of prostatitis and diffuse alveolar hemorrhage in the literature.

The diagnosis of Wegener granulomatosis is usually made by the histologic demonstration of three essential findings: small- and medium-vessel vasculitis, patchy or geographic necrosis, and granulomatous inflammation with neutrophils, lymphocytes, plasma cells, macrophages, giant cells, and eosinophils. The prostatic biopsy specimen obtained from our patient showed all of these features. The organ site and the amount of tissue obtained influence the likelihood of obtaining a positive biopsy result. The yield from upper airway biopsies is typically lower than those obtained from the lungs.

Life-threatening Wegener granulomatosis requires urgent treatment with high doses of glucocorticoids and cyclophosphamide. The medications are usually continued for at least 1 year after remission. For patients who have an exacerbation of the disease, long-term use of the least toxic drug for the maintenance of remission may be appropriate. This may include methotrexate, azathioprine, and low-dose prednisone.

Our patient presented initially with extrapulmonary manifestations, including prostatitis, transient
vision, and hearing loss followed by fulminant hypoxicemic respiratory failure secondary to diffuse alveolar hemorrhage. The initial differential diagnosis included bacterial, mycobacterial, and fungal pneumonia in addition to vasculitis. His treatment included antimicrobials to bacteria and tuberculosis and corticosteroids.

Bronchoscopic examination showed normal airways. The BAL revealed frank blood and a predominance of neutrophils. Results of bacterial, fungal, mycobacterial, and viral cultures and special stains were negative. Serum antinuclear and anti-glomerular basement membrane antibodies, and rheumatoid factor were negative. His tuberculin skin test was negative, and anti-neutrophil cytoplasm antibodies were positive. The clinical manifestation of hemoptysis, dyspnea, iron deficiency anemia, and a bilateral air space filling pattern on chest radiography were consistent with the syndrome of diffuse alveolar hemorrhage. Cyclophosphamide was added, and antimicrobials were discontinued. The patient improved rapidly over 14 days, was extubated, and was ultimately discharged breathing well on room air. He remained on cyclophosphamide and a reduced dose of corticosteroids. Figure 3 shows his chest radiograph on discharge from the hospital.

**Clinical Pearls**

1. The diagnosis of Wegener granulomatosis should be considered in patients who present with seemingly disparate multiorgan manifestations, especially when the respiratory, urogenital, ophthalmic, and otic systems are involved.
2. Symptomatic involvement of the lower genito-urinary tract in Wegener granulomatosis, although uncommon, may sometimes precede the onset of pulmonary disease.
3. Diffuse alveolar hemorrhage in Wegener granulomatosis is a life-threatening event that responds to steroids and cyclophosphamide.

**Suggested Readings**


**Figure 3.** The chest radiograph at the time of discharge showed impressive improvement of the lesions. Residual alveolar opacification still obscures much of the left hemidiaphragm. Alveolar and ground glass opacification are also present, especially in the right mid-lung. The costophrenic angles are blunted.