spaces and showed that it was the same as that expected in the trachea and was far greater than that expected in ambient air. The results of this test enabled us to determine with certainty that the source of the intraperitoneal gas causing tension pneumoperitoneum in our patient was from the lung, and an unnecessary laparotomy was avoided. The only gastrointestinal source that could practically cause a tension pneumoperitoneum with similar Po_2 measurements in gas specimens would be a tracheoesophageal fistula, but there was no evidence of gastrointestinal dilatation in our patient.

Regardless of the cause, the emergent treatment of tension pneumoperitoneum in an unstable patient should be percutaneous catheter insertion or, if time permits, drain placement to relieve the intraperitoneal pressure. This maneuver usually results in immediate hemodynamic and ventilatory improvement.* Thereafter, treatment depends on the specific cause. Although laparotomy may be necessary if a gastrointestinal cause is suspected, a surgical procedure should be avoided if the gas has dissected into the peritoneal cavity from the lungs. Patients with tension pneumoperitoneum due to pulmonary barotrauma may require continuous intraperitoneal catheter drainage for the duration of positive-pressure ventilation.

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Wegener's Granulomatosis Presenting as Dilated Cardiomyopathy

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WEGENER'S GRANULOMATOSIS is characterized by necrotizing, vasculitic, granulomatous lesions of the kidneys and upper and lower respiratory tract. Clinically notable cardiac involvement with the disease process is only rarely encountered. We describe the case of a patient who initially presented with dilated cardiomyopathy and who was later found to have elevated levels of antineutrophil cytoplasmic antibodies (c-ANCA); renal and pulmonary biopsy findings were consistent with Wegener's granulomatosis.

Report of a Case

The patient, a 57-year-old woman, presented with dyspnea on exertion and paroxysmal nocturnal dyspnea for two years. Eighteen months after her symptoms started, she was noted to have proteinuria and mild renal insufficiency. In July 1995, she underwent a cardiac evaluation

(Day JD, Ellison KE, Schnittger I, Perlroth MG: Wegener's granulomatosis presenting as dilated cardiomyopathy. West J Med 1996; 165:64-66)

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at an outside hospital for dyspnea on exertion after walking one block. An exercise stress test was stopped at stage I because she had severe shortness of breath. Dobutamine echocardiography showed global hypokinesis and four-chamber enlargement with possible apical and anteroapical ischemia. A subsequent coronary angiography was remarkable only for 30% to 40% stenosis of the proximal left anterior descending artery. A month later, she presented in pulmonary edema due to dilated cardiomyopathy and acute renal failure. At that time, her serum creatinine level was 430 μmol per liter (4.9 mg per dl), the hematocrit was 0.28 (27.9%), and a c-ANCA level was 18.6 equivalent units per ml (normal < 2.5). Her leukocyte count, creatine kinase, C3 complement, and C1 esterase levels were all within normal limits. A chest x-ray film showed cardiomegaly, pulmonary edema, and bilateral pleural effusions.

Echocardiography was repeated and was unchanged except for the new onset of a small pericardial effusion. A percutaneous renal biopsy showed crescentic glomerulonephritis without immune complex deposition, consistent with Wegener's granulomatosis. Of note, the patient had two episodes of supraventricular tachycardia, one described as multifocal atrial tachycardia and the other as atrial fibrillation, both of which responded to the intravenous administration of digoxin and diltiazem. She was discharged home on a regimen of diuretics, 1 mg per kg per day of oral cyclophosphamide, and 1 mg per kg per day of prednisone.

The patient did well at home for about a month until she presented to Stanford University Medical Center, Stanford, California, in September 1995 with increased dyspnea and worsening renal function. A chest radiograph showed cardiomegaly and focal areas of air-space disease throughout the right lung. The electrocardiogram was remarkable for left ventricular hypertrophy with repolarization abnormalities laterally. Echocardiography now showed worsened systolic function—ejection fraction, 0.20 (20%), as calculated by the ellipsoid-single plain method in the apical four-chamber view (Hewlett-Packard, Palo Alto, California, "Sonos Phased Array Imaging System Reference Guide," June 1994)—and moderate mitral regurgitation without any evidence of valvular vegetation. During this hospital stay, she was treated more aggressively for her Wegener's granulomatosis with intravenous cyclophosphamide, 2 mg per kg per day, and methylprednisolone sodium succinate. In addition, hemodialysis was started, with the placement of an arteriovenous fistula. A bronchoscopy with pulmonary biopsy was remarkable for grossly hemorrhagic alveoli, normal cytologic smears, and cultures negative for pathogens. She responded well to this regimen with a near return to normal mitral valve function, improved systolic function—ejection fraction, 0.30 (30%)—and pronounced symptomatic improvement.

Discussion

In this patient with Wegener's granulomatosis presenting as dilated cardiomyopathy, an endomyocardial biopsy was not done because of the low clinical yield of cardiac biopsies for diagnosing the cause of dilated cardiomyopathy.[1] Although histologic proof of myocardial involvement is lacking in this patient, the onset of congestive heart failure and renal involvement are temporally related. Moreover, global systolic function, as measured by echocardiography, appeared to decrease in conjunction with declining renal and pulmonary function after initial inadequate treatment of Wegener's granulomatosis. In addition to worsening global systolic function, mitral regurgitation developed in our patient, a finding not seen on two previous echocardiograms, which may be consistent with Wegener's granulomatosis-induced valvulitis or annular dilatation due to her dilated cardiomyopathy.[2-6] It is interesting to note that after more intensive therapy for Wegener's granulomatosis had been initiated, her systolic function improved and the mitral regurgitation nearly disappeared. This dramatic improvement of mitral valve function may be a result of treatment of the underlying Wegener's granulomatosis, as has been previously reported,[3] or of improved left ventricular function.

Since its initial description in 1931[7] and later confirmation in 1936,[8] Wegener's granulomatosis has been characterized histologically by necrotizing granulomatous vasculitis of the upper and lower respiratory tracts and the kidneys. Although the cause is unknown, the disease is generally considered a hypersensitivity disorder. It occurs most commonly in the fourth and fifth decades of life, with equal frequency among men and women. Wegener's granulomatosis usually develops over a 4- to 12-month period, with 90% of the patients presenting initially with upper or lower respiratory tract symptoms.[9-11] Although limited pulmonary forms of Wegener's granulomatosis have been described, renal involvement later in the disease process is nearly uniform.[12] The diagnosis of Wegener's granulomatosis in the past was made histopathologically, but the recent detection of characteristic circulating autoantibodies to neutrophil cytoplasmic antigens now permits earlier diagnosis and treatment.[13,14] Without treatment, the condition is usually fatal within a year of diagnosis. Despite the treatment-related morbidity, current therapy consisting of the use of glucocorticoids and cyclophosphamide allows complete remission in 75% of patients.[11]

In Wegener's initial paper describing the disease, he noted the possible involvement of the heart in the disease process.[8] Although subsequent autopsy studies have shown the heart to be involved frequently in Wegener's granulomatosis, clinical studies report the incidence of cardiac involvement as 8% to 12%.[10,11,15] The most common cardiac histopathologic findings reported are pericarditis and coronary arteritis in 50% of the patients, followed by focal myocarditis (25%), valvulitis or endocarditis (21%), conduction system defects (17%), and myocardial infarction (11%).[15]

In a 1980 histopathologic review of 27 case reports of cardiac involvement of Wegener's granulomatosis, 7 of the cases (26%) had evidence of focal myocarditis at autopsy.[15] Several reports exist where focal myocarditis led

to a clinical diagnosis of dilated cardiomyopathy. Of these rare reports, dilated cardiomyopathy was not found until after treatment with cyclophosphamide had been initiated.[4,10] In patients with bone marrow transplantation where much higher doses of cyclophosphamide are used, dilated cardiomyopathy may result from cyclophosphamide toxicity.[16,17] To our knowledge, this is the first case of Wegener's granulomatosis described in which the patient originally had dilated cardiomyopathy.

Unfortunately for our patient, even though the Wegener's granulomatosis was diagnosed, she was not adequately treated with corticosteroids and cyclophosphamide. Thus, over the month following diagnosis, her cardiac function declined further, manifested by a worsening ejection fraction and mitral regurgitation, extensive pulmonary involvement, and the need for lifelong hemodialysis. This case shows that although the clinical manifestation of cardiac disease in Wegener's granulomatosis is not usually encountered, it should be considered in the differential diagnosis of dilated cardiomyopathy in a patient with newly occurring pulmonary, renal, or other multisystem disorders.[18]

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