flow from severe aortic regurgitation during ventricular diastole.

In summary, an aneurysm of the left sinus of Valsalva, an unusual complication of aortic valvular endocarditis, was detected with two-dimensional echocardiography. Rupture of the aneurysm into the left atrium and associated aortic valvular dysfunction were demonstrated with pulsed Doppler echocardiography. These ultrasonic techniques provide complementary information and are useful in the diagnosis of endocarditis and its complications.

REFERENCES

Allergic Angitis of Churg and Strauss Syndrome*
Response to Pulse Methylprednisolone

R. MacFadyen, M.D.; V. Trom, M.D.; M. Keshmiri, M.D.; and J. D. Road, M.D.

*From the Department of Medicine, University of Saskatchewan, Saskatoon, and the Departments of Medicine and Pathology, University of British Columbia, Vancouver, Canada.
Reprint requests: Dr. Road, Department of Medicine, 2200 Westbrook Mall, Vancouver, B.C., Canada V6T 1W5

Our patient presented with widespread airspace consolidation. He was a steroid-dependent asthmatic receiving moderate doses of corticosteroid therapy. Open lung biopsy showed allergic angiitis of Churg and Strauss syndrome. The patient continued to deteriorate on high doses of prednisone. He was subsequently given four pulses of intravenous methylprednisolone with dramatic clearing noted on x-ray film and resolution of his shunt.

Churg and Strauss originally described the entity of allergic angiitis and granulomatosis in 1951. In 1977, Chumley et al reviewed 30 cases which fit the clinical and pathologic findings described by Churg and Strauss. They described eight cases with pulmonary involvement, and noted occasional complete regression with corticosteroid treatment. We describe a case of allergic angiitis and granulomatosis in which the patient presented with isolated severe pulmonary involvement even while on moderate doses of corticosteroids. The patient rapidly deteriorated until given pulses of methylprednisolone which resulted in prompt reversal of the disease.

CASE REPORT

The patient was a 20-year-old wheat farmer who presented with cough, wheezing and nocturnal dyspnea. A clinical diagnosis of asthma was made. Approximately five months later he presented with almost complete consolidation of the right upper lobe. At that time he had a peripheral eosinophil count of 6,000 per cu mm. His serum IgE was greater than 1,000 units. Skin tests for Aspergillus were negative. He did, however, have one precipitin line consistent with Aspergillus fumigatus, type 6. Results of roentgenograms of the sinuses, urinalysis and creatinine clearance were normal. The clinical diagnosis was eosinophilic pneumonia and he was treated with prednisone, 30 mg a day. One week later the infiltrate had cleared.

The patient was subsequently treated with prednisone in doses of 5-30 mg over the following 15 months to control symptoms due to asthma. No further parenchymal abnormalities were seen on chest

**Figure 1.** Anteroposterior chest roentgenogram showing widespread airspace consolidation predominantly in the lower lobes.
roentgenograms. His eosinophil count remained elevated (greater than 500/cu mm) throughout this period. Sodium cromoglycate was started 11 months after discharge from the hospital. Sixteen months after discharge, he presented with worsening cough, dyspnea and hypoxemia. He had been taking prednisone, 20 to 30 mg per day for the preceding five months. His eosinophil count was 140 cu mm on 30 mg a day of prednisone and he was febrile. The chest roentgenogram showed many areas of consolidation, predominantly in the lower lobes (Fig 1). The areas of consolidation were central rather than peripheral. The patient's steroid dose was increased to the equivalent of prednisone, 100 mg a day. Antibiotics were added to the regimen and an open lung biopsy was performed. Microscopically, the biopsy showed aggregates of eosinophils throughout and focally these collections were centered on blood vessels. Granulomatous and eosinophilic inflammation was demonstrated around tissue necrosis. Elastic stains (Fig 2), used to assess vessel destruction, showed disruption of the elastic lamina of arteries and veins; this feature confirmed the presence of a vasculitis. The finding, therefore, of a necrotizing vasculitis and a large number of eosinophils was consistent with allergic angiitis and granulomatosis (Churg-Strauss syndrome). After five days of steroid therapy equivalent to 100 mg of prednisone a day, his Po2 was 50 mm Hg on 15 liters of oxygen per minute by rebreathing mask and the infiltrates had worsened. Methylprednisolone was administered at a dose of 1 g a day for four days. After the first pulse, his eosinophil count fell to zero for the first time. His hypoxemia and pulmonary infiltrates then progressively improved. Six days later, on room air, his Po2 was 61 mm Hg and by ten days his chest roentgenogram had cleared dramatically. He was maintained on prednisone 60 mg a day and returned home. After one and a half years he has had one minor recurrence of the pulmonary infiltrates while on prednisone, 30 mg. This responded to a modest increase in prednisone dosage. Cyclophosphamide has since been added to attempt to reduce his steroid requirements. He continues to demonstrate no evidence of extrapulmonary involvement.

**Discussion**

This patient represents an unusual presentation of pulmonary infiltrates with eosinophilia. The initial presentation was suggestive of chronic eosinophilic pneumonia initially described by Carrington.14 Carrington's patients were mostly middle aged women who presented with a characteristic chest roentgenogram of peripheral areas of consolidation, the so-called "photographic negative of pulmonary edema." These patients responded very well to therapy with oral prednisone in doses of 20-40 mg per day. This patient is a young man whose roentgenographic presentation was of bronchocentric consolidation rather than peripheral areas of consolidation. The radiographic infiltrates of the allergic angiitis of Churg and Strauss syndrome have been described as transient patchy pneumonic infiltrates, massive bilateral nodular infiltrates without cavitation, and diffuse interstitial disease.1.5 The distribution of pulmonary infiltrates in our patient resembled that of the Churg-Strauss syndrome more than chronic eosinophilic pneumonia, but the degree of pneumonic consolidation was unusual. Although the pneumonitis responded to prednisone in the usual doses on the first admission, on the second admission the infiltrates developed while the patient was on moderate doses of steroid, which was very unusual for chronic eosinophilic pneumonia and more suggestive of an alternate diagnosis. The open lung biopsy specimen was compatible with the allergic angiitis of Churg and Strauss. These patients usually develop asthma before the onset of pulmonary vasculitis. The duration between the onset of asthma and onset of vasculitis is inversely correlated with the severity of the vasculitis.6 In this regard our patient would be expected to have a more severe course. Sodium cromoglycate has been reported as a possible cause of pulmonary infiltrates with eosinophilia.5.7 In our case, infiltrates preceded the use of sodium cromoglycate by one year, and the infiltrates worsened after it was stopped.

Active pulmonary involvement with the allergic angiitis of Churg and Strauss syndrome has been reported occasionally to completely regress on prednisone therapy.8 In this case, once the diagnosis was established by open lung biopsy, a clinical response was seen only after administering pulses of intravenous methylprednisolone. The patient might have improved with high-dose therapy with more time, but a rapid response was needed due to the patient's rapidly deteriorating condition. Methylprednisolone in high dose pulses has been reported to be effective in certain disease states,9,10 and particularly with fulminant pulmonary involvement with the vasculitis of Wegener's granulomatosis.10 Cyclophosphamide has also been advocated in this situation, but the response is often delayed for several weeks.11 The side effects of pulse corticosteroid therapy may include variations in blood pressure, as well as joint and CNS manifestations.11 In general, however, pulse therapy has few side effects.1 We are aware of no previous reports of the use of pulse methylprednisolone in the treatment of the allergic angiitis of Churg and Strauss syndrome. In this patient we believe the high dose pulse methylprednisolone prevented the need for assisted ventilation to maintain oxygenation and resulted in a dramatic improvement in the roentgenographic appearance.

In patients presenting with this clinical syndrome, a lung biopsy is mandatory before considering therapy. Large clinical trials to determine the ideal form of therapy will obviously be difficult due to the low incidence of this disease. However, the rapid response in this patient indicates this form of therapy may prove useful in treating the allergic angiitis of Churg and Strauss syndrome, particularly when a rapid response is required, and supports the findings of Chumbley et al12 who found occasional complete regression of pulmonary involvement with therapy with corticosteroids. This case also demonstrates that isolated pulmonary involvement can be a presenting feature of this disease.
Pseudoidiopathic Hypertrophic Subaortic Stenosis in a Patient with Cardiac Tamponade

A. Sundra Pandi, M.B.B.S.; and G. Kronik, M.D.

A 53-year-old woman with a large pericardial effusion and tamponade presented with signs of IHSS including a grade 4/6 apical systolic murmur, severe SAM, early systolic aortic valve closure and a small hypercontractile left ventricle but at most borderline left ventricular hypertrophy. Following pericardiocentesis, the clinical and echocardiographic signs of subvalvular obstruction resolved completely. One year later the patient died of bronchial carcinoma and no evidence of hypertrophic cardiomyopathy was found at autopsy. Pericardial tamponade should be added to the list of possible causes of dynamic subvalvular obstruction in a structurally normal heart.

Echocardiography is now the standard method for diagnosing idiopathic hypertrophic subaortic stenosis (IHSS). The three most important echocardiographic signs of this disease are: 1) asymmetric septal hypertrophy (ASH), 2) systolic anterior motion of the mitral echoes (SAM), and 3) mid-systolic closure of the aortic valve. While ASH is a sign of hypertrophic cardiomyopathy, SAM and premature aortic closure indicate dynamic subvalvular obstruction. However, none of these signs is totally specific for IHSS and in the literature there are occasional case reports on patients with dynamic subvalvular obstruction in the absence of hypertrophic cardiomyopathy. In this article, we report, for the first time, a patient in whom a large pericardial effusion with tamponade led to the appearance of clinical and echocardiographic signs of IHSS, which resolved completely after pericardiocentesis.

CASE REPORT

The patient was a 53-year-old woman who had been essentially healthy previously except for intermittent claudication of the left leg which was relieved by balloon dilatation of the femoral artery in 1979. On May 14, 1984, she was admitted to another hospital with progressive dyspnea, chest discomfort and fever (38.5°C). At that time, tachycardia, cardiomegaly and mild hepatomegaly were noted, but there was no systolic murmur. She was treated with antibiotics, digitalis and diuretics, yet deteriorated progressively. Three days later, a new grade 4/6 systolic murmur was noted at the apex and left parasternal area. She also developed bilateral pleural effusion, crural edema and hepatomegaly (4 cm below the costal margin). Bacterial endocarditis was suspected and the patient was transferred to our hospital. On arrival, she was sent to the echocardiography laboratory.

The echocardiogram (Fig 1) revealed no evidence of bacterial endocarditis. Instead, there was a large pericardial effusion with right ventricular compression as indirect evidence of cardiac tamponade. The left ventricle was small and hyperactive with an end-diastolic diameter (LVEDD) of 3.5 cm, an end-systolic diameter (LVEDS) of 1.5 cm (fractional shortening = 57 percent). The left ventricular outflow tract (LVOT) at the onset of systole was 1.6 cm, the septum 1.2 cm and posterior wall 1.2 cm. In addition, there was severe SAM of the mitral valve leaflets with broad contact to the interventricular septum. Using the method of Pollick et al., a subaortic gradient of 75 mm Hg was calculated. The aortic valve showed obvious midystolic closure with systolic fluttering. On the basis of these echocardiographic and clinical findings, the diagnosis of IHSS was made though there was no ASH and the left ventricular hypertrophy was unusually mild.

Since the patient was in very poor condition with tachycardia, pulsus paradoxus, low blood pressure, severe dyspnea and distended neck veins, pericardiocentesis was performed immediately and a pig tail catheter was introduced into the pericardial sac. After removal of 940 ml of clear fluid with high protein content, the systolic murmur became much softer. Serial follow up echocardiograms showed diminution and later disappearance of the echocardiographic signs of subvalvular obstruction with a simultaneous increase in the LVEDD, LVEDS and LVOT (Fig 2 and 3).

On the sixth day, the echocardiogram was normal except for some residual pericardial effusion and upper limit of normal left ventricular wall thickness (LVEDD = 3.8 cm, LVEDS = 2.2 cm, fractional shortening = 42 percent, posterior wall = 1.1 cm, ventricular septum = 1.1 cm). The phonocardiogram and pulse curve recorded at that time were normal at rest and during provocation with nitroglycerin. The patient progressively improved with supportive therapy and was discharged on the tenth day after complete recovery.

In July 1985 the patient was readmitted because of bronchial carcinoma and died shortly after admission. At autopsy the heart did not show any evidence of hypertrophic CMP

DISCUSSION

In this report, we describe a patient who developed prominent SAM and early closure of the aortic valve and a new and varying systolic murmur during the course of pericarditis with effusion and tamponade. These signs resolved completely after treatment of the pericardial disease.

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*From the First Department of Internal Medicine, Cardiology Division, University of Vienna, Austria.

Reprint requests: Dr. Kronik, Saarplatz 20, Vienna, Austria A-1090