Peripheral Pulmonary Embolization from Central Pulmonary Aneurysm*

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A 59-year-old man underwent successful repair of a pulmonary arterial aneurysm because of peripheral pulmonary embolization. These lesions are relatively rare; and, to our knowledge, peripheral embolization from such an aneurysm has not been previously reported.

Central pulmonary arterial aneurysms are rare lesions, occurring in one case per 13,696 autopsies, or 250 times less frequently than aortic aneurysms.1 Reports of successful repair are few and recommend repair in order to avoid the complications of rupture and right-sided heart failure or because of associated congenital lesions.2 Our case represents surgical repair of an isolated pulmonary arterial aneurysm because of peripheral pulmonary embolization.

CASE REPORT

A 59-year-old black man came to the emergency room on Oct 3, 1976, with symptoms of vague pain in the chest for one week and the recent onset of severe dyspnea, pressure-like pain in the chest, and a sense of impending doom. Pertinent physical findings were limited to the chest and were prolongation of expiration, expiratory wheezing, and a faint systolic murmur at the primary pulmonic area, with an increase in the pulmonary second sound.

On admission, the chest x-ray film (Fig 1) showed a large mass at the level of the pulmonary conus. Dissecting aneurysm was considered, but the aortogram was within normal limits. A pulmonary angiogram (Fig 2) showed the mass to be a large aneurysm of the main pulmonary artery. Right cardiac catheterization revealed normal right-sided pressures, with a mean of 23 mm Hg in the pulmonary artery and 33 mm Hg in the right ventricle. Right ventricular angiographic studies showed an area of radiolucency in the left main pulmonary artery and decreased perfusion of the right lower lobe.

Surgical repair was planned because of the possibility of rupture and because of the impression that the patient was having recurrent pulmonary emboli from the aneurysm. On opening the pericardium, a large fusiform aneurysm extended from the pulmonary annulus to the origins of both branches of the pulmonary artery. The patient was placed on cardiopulmonary bypass, and the aneurysm was opened longitudinally. Multiple small thrombi (under 6 mm) were obtained from the distal left circulation with a balloon-tipped catheter. Then the aneurysm was plicated to produce a pulmonary artery of normal caliber.

Despite a slightly prolonged postoperative stay due to pulmonary infection, the patient did well and was discharged about two weeks after surgery. Pathologic examination of the resected specimen showed mucoid degeneration of the media and organized thrombi from the distal circulation. The patient has done well since surgical repair.

DISCUSSION

Two early articles1,2 present series of autopsies with detailed reviews of the literature. More recent case reports have discussed the association of pulmonary arterial aneurysm with congenital heart disease,3-6 with these
cases including about 45 percent of all such lesions.1-3

Diagnosis is generally made on the basis of an abnormal chest x-ray film in a patient with exertional dyspnea, cough, and pain in the chest. Right cardiac catheterization with pulmonary angiographic studies delineates the lesion and rules out associated congenital heart disease. Death in untreated patients is generally due to rupture; the average age of untreated patients at death is about 40 years.4

This case represents peripheral embolization from a central pulmonary arterial aneurysm. Although this clinical situation is well described with aneurysms in the systemic circulation, it has not been previously described in the pulmonary vascular tree. The possibility of embolization should enter into the decision for repair of such a lesion.

CONCLUSION

Pulmonary arterial aneurysms will be found more frequently with modern techniques of angiography. This report describes successful repair of such a lesion for a hitherto unreported complication.

REFERENCES


Acute Respiratory Failure and Pulmonary Arteritis Without Parenchymal Involvement*

Demonstration in a Patient With Rheumatoid Arthritis

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A 28-year-old woman with an eight-year history of rheumatoid arthritis presented with a three-day history of dyspnea. Physical and electrocardiographic findings were consistent with pulmonary hypertension. Arterial blood gases revealed a ventilation-perfusion mismatch. Chest roentgenogram was normal. After transient improvement, she suddenly deteriorated and died. At autopsy, a necrotizing pulmonary panarteritis was found without parenchymal involvement by rheumatoid disease. The pulmonary arteries were the only vessels affected. Immunofluorescent staining revealed immunoproteins scattered throughout the vessel walls without localization to the basement membrane. The unique features of the case are discussed in relation to pulmonary hypertension and rheumatoid lung disease in which vascular lesions are usually associated with honeycomb lung. The association between the rheumatoid arthritis and pulmonary vasculitis was probably coincidental.

Pulmonary arteritis is a rare complication of rheumatoid arthritis. When it occurs, it usually occurs with parenchymal lung disease such as fibrosing alveolitis or honeycomb lung, and cor pulmonale.1 There are two cases reported in the English literature of primary rheumatoid pulmonary vasculitis without underlying parenchymal disease. Of these two cases, ours is the first patient who developed acute respiratory failure and died.

CASE REPORT

History and Clinical Course

A 28-year-old waitress was admitted to the Rheumatology Service of Rancho Los Amigos Hospital on Sept 19, 1977, for dyspnea of three days' duration.

The patient developed rheumatoid arthritis in 1970. Disability due to multiple joint involvement progressed despite several courses of salicylates, indomethacin, and phenylbutazone prescribed by private physicians. She was referred to the Rancho Los Amigos Hospital Rheumatology Service in April 1977 for decreasing hand function. Physical and roentgenographic examinations demonstrated extensive rheumatoid arthritis changes. Latex fixation titer was 1:320 and antinuclear antibody titer 1:640. There was no evidence of iritis. Felty's syndrome, Raynaud's phenomenon, or cutaneous vasculitis. Chest roentgenogram was normal. The patient was placed on salicylates, 2.9 gm, and prednisone, 8 mg/day, and a vigorous physical therapy program. The proximal interphalangeal joints were surgically fused in the right hand with improvement of hand function.

The patient did well at home until three days prior to the final admission when she noted exertional dyspnea. She did not smoke or take contraceptive medication or illicit drugs. Physical examination revealed a cushingoid woman in respiratory distress. Temperature was 37.8°C, blood pressure, 90/60 mm Hg, pulse rate, 100 beats per minute and regular, and respirations, 24 per minute. There was no neck vein distention, hepatojugular reflux, or hepatomegaly. Lungs were normal upon auscultation and percussion. There was an intermittent gallop rhythm and increased intensity of the pulmonic sound. Extremities revealed extensive joint deformities but no acutely inflamed joints or phlebitis. There