histologic confirmation of the diagnosis and postoperative follow-up studies. These studies indicate that MRI can detect central pulmonary emboli. They suggest that it may be less useful in the detection of peripheral emboli.

One of the potential problems in using MRI to evaluate suspected pulmonary embolism is the differentiation of thrombus from pulmonary arterial hypertension. In the setting of pulmonary arterial hypertension, slow flow within the pulmonary arteries can produce intraluminal signal. A recent study by White et al. using cardiac gated MRI addressed this problem of differentiating pulmonary embolus from pulmonary arterial hypertension. Their investigations suggested that thrombus appears as a discrete intraluminal signal which remains constant throughout the cardiac cycle and exhibits decreased signal intensity on the second echo image relative to the first echo image. Pulmonary arterial hypertension without thromboembolism produces an intraluminal signal which appears to change in distribution during the cardiac cycle, disappears at one phase of the cycle, and exhibits increased signal intensity on the second echo image. The surgically confirmed thromboembolus in our patient behaved on MRI precisely as suggested by White et al. with a signal constant in distribution and extent throughout the cardiac cycle and decreasing in intensity from first to second echo.

Pulmonary embolism is an uncommon cause of unilateral absence of perfusion or unilateral massive perfusion defect on pulmonary perfusion scintigraphy. Nevertheless, many patients with these scintigraphic findings undergo pulmonary angiography to rule out pulmonary embolus. Since MRI can detect not only central pulmonary artery thromboembolus but also other causes of massive unilateral perfusion defects such as bronchogenic carcinoma and mediastinal or hilar adenopathy, it may be a useful noninvasive alternative to angiography in evaluating these patients.

This case illustrates the potential utility of cardiac gated MRI in the evaluation and follow-up of central pulmonary artery thromboembolism.

REFERENCES
4. Thrickman D, Kressel HY, Axel L. Demonstration of pulmonary embolism by magnetic resonance imaging. AJR 1984; 142:921-22

Severe Pulmonary Hypertension with Diffuse Smooth Muscle Proliferation of the Lungs*

Pulmonary Tuberous Sclerosis?

Osvaldo E. Wagen, M.D.;† Aquiles J. Roncoroni, M.D., F.C.C.P.;‡ and Juan A. Barcat, M.D.§

A 49-year-old man with normal intelligence and no cutaneous lesions had symptoms and signs of cor pulmonale. The chest roentgenogram showed an interstitial pattern. Hemodynamic studies revealed increased pulmonary arterial and wedge pressures. The patient died in shock, and the postmortem findings were a diffuse smooth muscle proliferation in the lungs as the cause of the pulmonary hypertension. We presume that this is a form of pulmonary tuberous sclerosis of which few cases are reported with such severe pulmonary hypertension as shown by our measurements. (Chest 1989; 93:234-37)

Diffuse smooth muscle proliferation involving alveolar, bronchiolar, and vessel walls and pulmonary interstitium is uncommon. Since the report of Berg and Vejlens, it has been recognized that such proliferation may represent the pulmonary component of tuberous sclerosis, a disease characterized by epilepsy, mental retardation, and adenoma sebaceum. Similar pulmonary lesions may be found in patients without other evidence of this last disease. Muscle proliferation may affect intrapulmonary lymphatic vessels, mediastinal lymph nodes, and the thoracic duct, producing chylous effusions in pulmonary lymphangiomatosis. In the absence of lymphatic involvement, the condition is named diffuse pulmonary leiomyomatosis. Many cases of diffuse pulmonary leiomyomatosis show one or more signs of tuberous sclerosis, a fact which induced Valent‡ to propose that diffuse pulmonary leiomyomatosis may represent a "forme fruste" of the disease.

The present report details the findings in a man without clinical or family history of tuberous sclerosis who had severe cor pulmonale and elevated pulmonary and wedge pressures which induced his death.

CASE REPORT

The patient was a 49-year-old man, a physician, referred because of dyspnea on minimum exertion. Symptoms started four years prior to admission with exertional dyspnea. Three years later, the patient developed dyspnea on mild exertion, edema of the lower extremities, and ascites. He received digoxin and diuretics, with symptomatic relief. An echocardiogram showed enlarged right-sided chambers. Two months later, the patient complained of cough and sudden dyspnea. With a presumption of pulmonary thromboembolism, he was treated with anticoagulants. Two months prior to admission, dyspnea on minimum exertion appeared. The patient had smoked 40 cigarettes daily for 27 years and met clinical criteria for severe pulmonary hypertension (Wagen, Roncoroni, Barcat)

[From the Instituto de Investigaciones Médicas "Alfredo Lanari," Facultad de Medicina, Universidad de Buenos Aires, Buenos Aires, Argentina.
†Chief Medical Resident.
‡Professor of Medicine and Medical Director.
§Chief Pathologist.
Reprint requests: Dr. Roncoroni, Ac. Donato Alvarez 3150, Buenos Aires, Argentina 1427]
for chronic bronchitis. He denied hemoptysis, chest pain, arterial hypertension, and cutaneous lesions, as well as any pathologic family history, including mental deficits.

The patient was a well-nourished man, with marked cyanosis of the skin and mucous membranes. The pulse rate was 88 beats per minute, arterial blood pressure was 85/50 mm Hg, axillary temperature was 36°C, and the respiratory rate was 20/minute. The patient had clubbing of the fingers, moderate edema of the lower limbs and sacrum, and jugular venous distention at a 45° angle, without inspiratory collapse. He had Dressler’s sign, an increased second heart sound, and a systolic murmur (+ + +) at four areas. The lung had crepitant rales at the right base. The patient had mild ascites and hepatomegaly without splenomegaly. The kidneys were not palpable. Findings from the neurologic and funduscopic evaluations were normal. There were no cutaneous lesions.

Laboratory data were as follows: hemocrit, 49 percent; leucocyte count, 7,300/cu mm with a normal differential cell count; blood urea nitrogen, 18 mg/dl; and creatinine clearance, 42 ml/min. The level of serum glutamic-oxaloacetic transaminase (SGOT) was raised fourfold, the level of serum glutamic-pyruvic transaminase (SGPT) was raised twofold, and the creatine kinase level was normal. Total bilirubin level was 3.85 mg/dl, and direct bilirubin level was 2.75 mg/dl. The plasma concentration of sodium was 135 mEq/L and of potassium was 4.9 mEq/L. The chest roentgenogram (Fig 1) showed normal pulmonary size, an interstitial pattern with "honeycombing." Kerley’s B lines were in both lower fields, cardiomegaly, and a prominent left middle arch and pulmonary artery. The electrocardiogram showed sinus rhythm, first-degree atrioventricular block, biventricular enlargement, clockwise rotation of the heart on frontal and horizontal planes, and right ventricular hypertrophy. Spirometry showed the following data: vital capacity (VC), 3.75 L (72 percent of predicted); forced expiratory volume in one second (FEV₁), 2.16 L (53 percent); and mean forced expiratory flow over the middle half of the forced vital capacity (FEF 25-75%) 1.08 L (25 percent). No changes were provoked by bronchodilators. Arterial blood gas levels while breathing room air at rest were as follows: arterial oxygen pressure (Pao₂), 47 mm Hg; pH, 7.57; arterial carbon dioxide tension (PaCO₂), 28 mm Hg; while breathing 100 percent oxygen, the PaO₂ was 355 mm Hg, and the arterial-alveolar oxygen ratio was 0.52. A Swan-Ganz catheter registered severe pulmonary hypertension with increased wedge pressure (Table 1).

Therapy with dopamine was started, and different vasodilators were tried, but the patient developed hypotension, remaining dopamine-dependent. Three days later, he suffered a cardiopulmonary arrest.

Necropsy was limited to the thoracic organs. The heart was enlarged and dilated. The weights were as follows: total, 512 g; right ventricle, 169 g, and left ventricle plus septum, 165 g. There were no valvular lesions; myocardial fibers of the right-sided chambers were hypertrophic. Coronary arteries showed a few fatty streaks in the intima. The pulmonary trunk and main arteries had fatty streaks and fibrous plaques; no thrombi were found. Mediastinal lymph nodes were up to 2 cm in diameter; the enlargement was due to diffuse lymphoid hyperplasia. Both lungs showed panacinar and bullous emphysema, predominantly in the upper lobes. Microscopically, the most important finding was a focal and disorderly proliferation of the smooth muscle in the pleura, septa, and bronchiolar and alveolar walls (including those of some emphysematous acini and bullae), interstitium, and blood vessel walls, especially in veins less than 1 mm in diameter (Fig 2 and 3). Some of the foci of muscle proliferation formed small nodules up to 4 mm in diameter. There was no interstitial fibrosis. Elastic and muscular arteries showed focal, eccentric fibrous patches in the intima (Fig 2); muscular arteries also showed medial hypertrophy. Intrapulmonary or extrapulmonary lymphatic vessels were not involved.

| Table 1 — Hemodynamic Monitoring with and without Vasoactive Drugs on Day 2 |
|--------------------------|-----------------|-----------------|----------------|-----------------|-----------------|
| Heart rate, beats per min | 3PM  | 3:45PM | 4:30PM | 5PM  | 11PM | 11:50PM |
| Mean arterial pressure, mm Hg | 90   | 90   | 96   | 75   | 70   | 70   |
| Mean pulmonary arterial pressure, mm Hg | 74   | 63   | 73   | 73   | 76   | 83   |
| Pulmonary arterial wedge pressure, mm Hg | 30   | 20   | 19   | 21   |     |     |
| Central venous pressure, mm Hg | 21   | 20   | 15   | 10   | 12   | 20   |
| Cardiac index, L/min/m² | 2.2  | 3.2  | 4.1  | 2.8  | 3.5  | 2.7  |
| Systemic vascular resistance, dynes·cm⁻⁵ | 1,346 | 965  | 859  | 1,000 | 730  | 817  |
| Pulmonary vascular resistance, dynes·cm⁻⁵ | 858  | 593  | 572  | 800  |     |     |
| FIo₂ | 0.21 |     |     |     | 0.40 |     |
| PaO₂, mm Hg | 47   |     |     |     | 62   |     |
| pH | 7.57 |     |     |     | 7.49 |     |
| PaCO₂, mm Hg | 28   |     |     |     | 28.5 |     |
| PvO₂, mm Hg | 27   |     |     |     |     |     |
| Dopamine, µg/kg/min |     | 10   | 10   | 10   | 15   |     |
| Sodium nitroprusside, µg/min |     | 100  |     |     |     |     |
| Captopril, mg |     |     |     |     |     | 6.5  |

*Fractional concentration of oxygen in inspired gas.
FIGURE 2. Smooth muscle masses surrounding bronchiole and small pulmonary artery. Artery also shows eccentric fibrous patch in intima (Verhoeff’s elastic stain, original magnification × 25).

The smooth muscle cells were well differentiated and without mitosis. Bronchi showed no signs of chronic bronchitis.

DISCUSSION

The main feature of the patient was the severe precapillary and postcapillary pulmonary hypertension and cor pulmonale. The diagnosis before hemodynamic studies was primary pulmonary hypertension or chronic silent recurrent thromboembolism. Pulmonary veno-occlusive disease or fibrosing mediastinitis were considered after high pulmonary wedge pressure was found. The state of the patient and his rapid death precluded further studies.

Although hemodynamic studies are sparse in pulmonary lymphangiomymomatosis and pulmonary tuberous sclerosis, one case of the latter had a mean pulmonary arterial pressure of 35 mm Hg at rest.4 Clinical or pathologic signs of right cardiac strain were also seen in five of 28 cases of pulmonary lymphangiomymomatosis.5 In that study, pulmonary arterial and venous walls showed thickening by inordinate muscle growth, reaching the point of venous occlusion. In our patient the increased gradient between pulmonary arterial diastolic and wedge pressures indicates an elevated arteriolar resistance,7 probably due to partial obstruction of the pulmonary arterial bed by “in situ” thrombi, hypoxic vasoconstriction, or the smooth muscle proliferation. High wedge pressure may reflect the increased resistance in the pulmonary venous bed or an elevated pressure in the left cardiac chamber. Wedge pressure may be equivalent to pulmonary capillary or left atrial pressure, depending on the position of the catheter’s tip in the pulmonary vascular bed.7 If it is in a large pulmonary artery, the junction point (the point in the venous side of pulmonary circulation in which the blood of the occluded vessel contacts with the blood of the nonoccluded) is located in a pulmonary vein about the same size as the occluded pulmonary artery, near the left atrium. If the tip is placed in a small artery, the junction point approximates that of the capillary bed, representing with greater accuracy its changes in resistance. In our patient the pressures were registered from the main branches of the pulmonary artery, so the values obtained should be equivalent to those proximal to the left atrium. The lack of lesions in the large pulmonary veins, left atrium, and mitral valve led us to think that the wedge pressure represented the end-diastolic left ventricular filling pressure, elevated because of a distorted left ventricular geometry and decreased distensibility by the large right ventricular volume and displacement of the septum towards the left chamber.4 No anatomic cause of left ventricular failure was found at necropsy.

Moderate ventilatory obstruction could be explained by muscle proliferation in the bronchiolar walls, with air-trapping.

Arterial hypoxemia in pulmonary vascular occlusion was attributed in the past to decreased transit time produced by a restricted vascular bed.4 Modern methods show that hypoxemia may be secondary to a variable, but rather small, shunt effect, decrease in mixed-venous oxygen pressure, and mainly ventilation-perfusion mismatch.10 All of these abnormalities were present in our patient; the increase in arterial-alveolar oxygen ratio observed under oxygen may be a measure of ventilation-perfusion inequality, mixed-venous oxygen pressure was 27 mm Hg, and shunt while breathing 100 percent oxygen, was 13 percent of cardiac output. The persistence and opening of a foramen ovale or any other intracardiac right-to-left shunt was excluded by the autopsy.

The reticular-interstitial pattern in the chest x-ray film may have been an expression of muscle overgrowth combined with interstitial edema, due to venous occlusion and/or left ventricular “dysfunction” due to right-left ventricle interaction. The most frequent radiologic changes in pulmonary tuberous sclerosis and diffuse pulmonary leiomyomatosis are reticular and motting patterns with honeycombing,9 generally in a diffuse form, but sometimes localized.

The main difficulty in the diagnosis of this patient is his sex. Both pulmonary lymphangiomymomatosis and pulmonary tuberous sclerosis (including the “forme fruste” diffuse pulmonary leiomyomatosis) are distinctly a woman’s disease, but two cases of pulmonary tuberous sclerosis have been described in men.4 Furthermore, in this case, the lack of smooth muscle proliferation with formation of sinuses or cysts in lymph nodes,11 which is a characteristic of pulmonary lymphangiomymomatosis,10 would suggest that the patient had pulmonary tuberous sclerosis. Pulmonary compromise in tuberous sclerosis is infrequent. It affects almost exclusively women, the average age of onset of symptoms of pulmonary...

Figure 3. Small vessels going through masses of smooth muscle (Verhoeff’s elastic stain, original magnification × 10).
involvement is 34 years, and generally the patients are of normal intelligence. Progressive dyspnea and nonproductive cough are the most frequent symptoms; less frequent are hemoptysis and clubbing. Our patient was a 49-year-old man with normal intelligence and dyspnea of slow progression. It is not valid to exclude tuberculous sclerosis for absence of a family history or lack of signs or symptoms of other organ involvement, since it may be sporadic in 75 percent of the cases, cerebral lesions can be asymptomatic, and adenoma sebacum may be absent in 15 percent of the cases. Moreover, in the review by Harris et al., one of the male patients with pulmonary tuberculous sclerosis had only pulmonary involvement, so it is not possible to exclude that condition as the final diagnosis.

The clinical and hemodynamic severity of pulmonary vascular involvement present in this case has not, to our knowledge, been reported yet. It seems that pulmonary tuberculous sclerosis and diffuse pulmonary leiomyomatosis should be added to the clinical roster as a rare cause of pulmonary vascular hypertension and right-sided heart failure.

REFERENCES

Recently, a new ECG lead has been introduced that can mimic a pulmonary nodule to the uninitiated. Alternatively, in those institutions where the lead is commonly seen, physicians may dismiss a real finding on the chest x-ray film as a lead "artifact." We describe the appearance of this new chest wall artifact. (Chest 1989; 95:237-238)

Artifacts are frequently present on chest roentgenograms, and the experienced observer can recognize most; however, as the number and variety of monitoring and therapeutic devices steadily proliferate, the number and variety of associated artifacts also increase. Observers may become familiar with artifacts generated by certain types of these devices. Unfortunately, different departments in many large hospitals will often use and purchase different brands, and new or uncommon artifacts may go unrecognized and be misinterpreted.

CASE REPORT

A 77-year-old woman came to the Emergency Department with severe abdominal pain. Her abdominal x-ray film was normal, but her frontal chest x-ray film demonstrated an ill-defined 1.8-cm pulmonary nodule in the left apex. The nodule could not be seen on the lateral chest x-ray film, but the apices of the lungs are typically difficult to evaluate on lateral views. Except for moderate cardiovascular findings due to hypertensive disease, no other cardiopulmonary abnormalities were seen. A thoracic CT scan showed no evidence for a pulmonary nodule or mediastinal abnormalities. The initial chest x-ray films were reevaluated, and it was determined that the nodule was the typical artifact produced by the ECG lead.

The patient therefore underwent an unnecessary thoracic CT scan. Theoretical risks include reaction to the contrast medium and

Pulmonary Nodule Mimicked by ECG Lead Artifact*

Tony Kim, M.D.; Richard N. Messersmith, M.D.; and Heber MacMahon, M.D., F.C.C.P.

*From the Department of Radiology, University of Chicago, Chicago.

Reprint requests: Dr. Kim, Department of Radiology, Box 429, 5841 South Maryland, Chicago 60637

FIGURE 1. Radiographic appearance of several ECG leads; offset gel can be confused with a pulmonary nodule.