Right Heart Failure Secondary to Compression of the Right Pulmonary Artery by a Syphilitic Aortic Aneurysm*

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A 68-year-old man presented with right heart failure due to compression of the right pulmonary artery by a syphilitic aneurysm of the ascending aorta. The diagnosis was made by cardiac catheterization and angiography and it was proven by autopsy. This complication is unusual and supports the experimental evidence that unilateral pulmonary obstruction may be responsible for the development of pulmonary hypertension.

Compression of the main pulmonary artery or its branches by an aneurysm of the ascending aorta is a rare complication.[1-5] Brill and Jones,[1] in 1946, stated that of the 87 reported cases of compression of the pulmonary artery by a syphilitic aortic aneurysm, 84 patients died from rupture of the aneurysm into the pulmonary artery and three died from cor pulmonale without rupture. Since studies have shown that unilateral pneumonectomy does not produce pulmonary hypertension,[6] it is of interest that unilateral compression of a pulmonary artery can produce pulmonary hypertension and right heart failure. The purpose of this article is to present a case of this complication diagnosed by cardiac catheterization and angiography and also proved at autopsy, and to discuss the mechanism of development of pulmonary hypertension under such settings.

Case Report

A 68-year-old black man was admitted to Hahnemann Medical College and Hospital in October, 1970, because of nonspecific chest pain, dyspnea on exertion and swelling of the legs for two weeks. Past medical history was unremarkable.

On physical examination, the heart rate was 100 beats per minute and irregularly irregular; his blood pressure was 140/40 mm Hg in both arms, and his temperature was 98.6°F (37°C). The carotid pulses were full, with a brisk upstroke. The neck veins were distended at 45°. A prominent left ventricular impulse was palpable in the sixth intercostal space at the anterior axillary line. The first and second heart sounds were normal. There was a grade 3/6 early decrescendo, diastolic blowing murmur heard equally well in both the right and left sternal borders, and a harsh grade 2/6 mid-systolic ejection murmur along the left sternal border radiating toward the base of the heart. The lungs were clear to auscultation. The liver was enlarged, palpable 10 cm below the right costal margin. There was 3+ pitting edema in both legs.

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Table 1—*Cardiac Catheterization Data

Left atrium	(12) V = 14
Pulmonary artery	79/22 (40)
Right ventricle	78/10
Right atrium	(8)
Cardiac output	3.6 L/min
Cardiac index	2.2 L/min/sq meter
Pulmonary vascular resistance	622 dynes sec cm^{-5}
Brachial artery	136/34 (72)
Ascending aorta	110/32 (66)
Left ventricle	116/11
Arteriovenous O_2 difference	6.0 Vol%
Systemic O_2 saturation	95%
Heart rate	70 beats/min

*Pressures are in mm Hg; mean pressure in parentheses.

The VDRL (Venereal Disease Research Laboratory) and FTA (fluorescent treponemal antibody) tests were positive. The chest roentgenogram showed a large calcified mass in the anterior mediastinum with an enlarged cardiac silhouette, mainly because of left ventricular enlargement. The electrocardiogram showed atrial fibrillation, with ventricular premature beats and deep "S" waves in the lateral precordial leads. The QRS vector loop suggested right ventricular hypertrophy.

Combined left and right heart catheterization was performed. The right heart catheter was advanced without difficulty into the main and the left pulmonary arteries; however, it was impossible to advance it into the right pulmonary artery. The hemodynamic data (Table 1) indicated normal left ventricular end-diastolic pressure and mean left atrial pressure; elevated pulmonary artery and right ventricular pressures; and increased pulmonary vascular resistance.

Aortography delineated a large saccular aneurysm of the ascending aorta (Fig 1). There was 3+ (maximum 4) aortic valve regurgitation. Left ventriculography showed a mildly enlarged left ventricular cavity, with a normal contraction pattern. On fluoroscopic examination, the large aneurysm was seen to be heavily calcified. Main pulmonary artery cineangiography showed good opacification of the main and the left pulmonary artery and its branches, and only trivial flow into the right pulmonary artery (Fig 2).

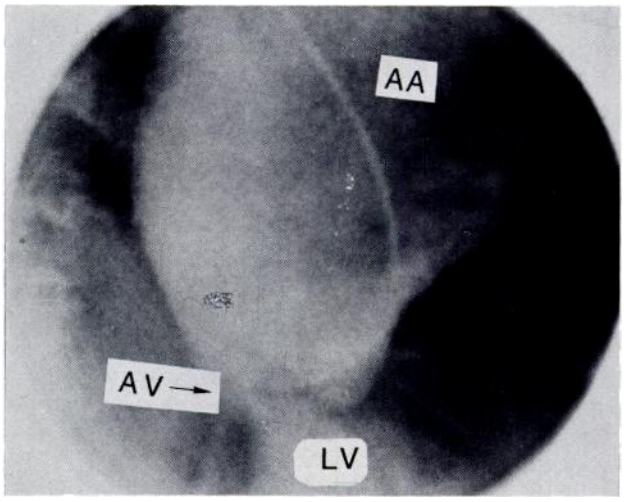

Figure 1. Ascending aortography in the right anterior oblique projection. Significant aortic valve insufficiency *(arrow)* and the large aneurysm of the ascending aorta are seen (AA). LV = left ventricle; AV = aortic valve.

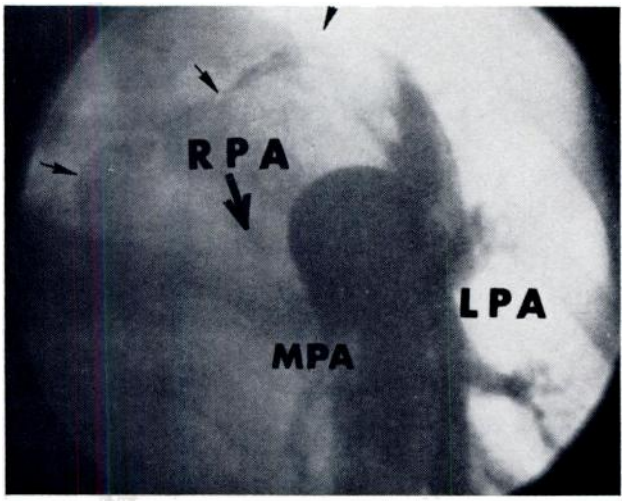

FIGURE 2. Pulmonary angiogram in slight right anterior oblique projection. The main pulmonary artery (MPA) and the left pulmonary artery (LPA) are well visualized. The *arrow* points to the area of the right pulmonary artery (RPA) which is not seen in this frame. Small *arrows* outline the calcified aneurysm.

Because of the patient's age and the high risk in inserting an aortic valve prosthesis with resection of the ascending aorta, surgery was not recommended, and he was discharged on a medical regimen.

He remained symptomatic with dominant right heart failure, poorly responsive to therapy. Four years later, he succumbed after an episode of ventricular fibrillation.

Autopsy Findings

The significant autopsy findings were limited to the cardiovascular system. A large saccular aneurysm of the ascending aorta measured 10 x 9 x 8 cm. The microscopic findings were consistent with chronic syphilitic aortitis. The right pulmonary artery was compressed by the aneurysm, and its orifice was probe-patent. The left pulmonary artery was normal and the main pulmonary artery was slightly dilated and thickened. The heart weighed 875 gm (including the aneurysm) and showed marked left ventricular dilatation and hypertrophy (wall thickness, 3 cm). The right ventricle was dilated and thickened (1.2 cm). The ostium of the left coronary artery was displaced by the aneurysm, but there was no narrowing, and the coronary arteries were free of significant atherosclerotic changes. There was no evidence of old or recent infarction. On microscopic examination, there was marked thickening and hyperplasia of the wall of the pulmonary arterioles (Fig 3).

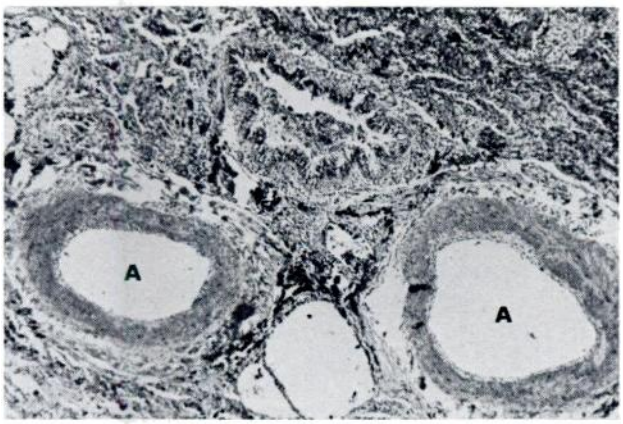

FIGURE 3. Microscopic section of the left lung showing marked thickening of the pulmonary arteriolar wall (Hematoxylin and eosin, original magnification × 40).

DISCUSSION

Compression of the main pulmonary artery or its branches is an unusual complication of syphilitic aneurysm of the ascending aorta[1-5] and only a few such cases have been studied by cardiac catheterization.[2-5] Our patient presented with right heart failure and pulmonary hypertension proven at cardiac catheterization. Although there was clinical and angiographic evidence of aortic valve disease, we do not feel that the pulmonary hypertension was secondary to left heart failure because the left atrial pressure and the left ventricular end-diastolic pressures were normal and the patient had no clinical evidence of left heart failure at any time. Moreover, angiographic and autopsy findings leave no doubt that the pulmonary hypertension was caused by severe compression of the right pulmonary artery.

This case is most interesting since it demonstrates that unilateral pulmonary artery obstruction can cause pulmonary hypertension despite the accepted fact that unilateral pneumonectomy does not cause significant elevation of the pulmonary arterial pressure.[6] There are several experimental and clinical supportive evidences which may shed light on the mechanisms in such cases.[7-9] Pool et al,[8] in an extensive review of congenital unilateral absence of a pulmonary artery, found that 19 percent of the patients without associated cardiac lesions have pulmonary hypertension. On the other hand, they have found that 88 percent of the patients with associated lesions and left-to-right shunt have pulmonary hypertension. Recently, Cohn and associates[10] described a 48-year-old patient with congenital right pulmonary artery stenosis who developed pulmonary hypertension. Experimental ligation of the pulmonary artery in dogs produces pulmonary hypertension in 50 percent of the animals if the ligation is performed on the first day of life.[7] However, if the ligation is done at several weeks of age, no pulmonary hypertension was observed.[7] Vogel et al[9] have found that ligation of the left pulmonary artery in calves fails to produce pulmonary hypertension if performed at sea level. If ligation is done at 5,200 feet above sea level, pulmonary hypertension was observed, presumably because the effect of hypoxia is additive to that of increased flow in the uninvolved lung. Moreover, since the right lung has a larger capacity than the left (55 percent versus 45 percent) Vogel and associates[9] have found that ligation of the right pulmonary artery even at sea level produced hypertension in the newborn calves, probably because the increased flow into the left lung in such cases is of sufficient magnitude to produce the vascular changes. From the above discussion, it is evident that the amount of blood flow delivered to the contralateral lung, the age at the onset of obstruction and the effect of hypoxia are important factors in the genesis of pulmonary hypertension in the presence of unilateral pul-

monary artery obstruction. Early diagnosis and improved surgical facilities should improve the prognosis in patients with similar but certainly uncommon clinical entity.

References

1 Brill IC, Jones RS: Syndrome of compression of the pulmonary artery by syphilitic aneurysm with and without arterio-arterial communication. Ann Intern Med 24:111, 1946

2 Donnel JL, Levinson DC, Griffith GC: Clinical studies of involvement of the pulmonary artery by syphilitic aortic aneurysm. Circulation 13:75, 1956

3 Jacoub MH, Bainbridge MV, Gold RC: Aneurysm of the ascending aorta presenting with pulmonary stenosis. Thorax 21:236, 1966

4 Bevis AG, Rojas RH, Stansel H: Aneurysm of the ascending aorta causing obstruction of the left pulmonary artery. J Thorac Surg 52:245, 1966

5 Watanabe T, Miyazawa K, Ksukamoto M, et al: Syphilitic aneurysm of ascending aorta producing pulmonary stenosis by compression. Am J Cardiol 20:575, 1967

6 Cournand A, Himmelstein A, Riley RL, et al: A follow-up study of the cardiopulmonary function in four young individuals after pneumonectomy. J Thorac Surg 16:30, 1947

7 Marshall RJ, Wang Y, Semler H, et al: Flow, pressure and volume relationship in the pulmonary circulation during exercise in normal dogs and dogs with divided left pulmonary artery. Circulation Res 9:53, 1961

8 Pool PE, Vogel JHK, Blount SG: Congenital unilateral absence of a pulmonary artery, the importance of flow in pulmonary hypertension. Am J Cardiol 10:706, 1962

9 Vogel JHK, McNamara DG, Hallman G, et al: Effects of mild chronic hypoxia on the pulmonary circulation in calves with reactive pulmonary hypertension. Circulation Res 21:661, 1967

10 Cohn LH, Sanders JH, Collins JJ: Surgical treatment of congenital unilateral pulmonary arterial stenosis with contralateral pulmonary hypertension. Am J Cardiol 38:257, 1976

Pericardial Tamponade Secondary to Sudden Steroid Withdrawal in Chronic Rheumatoid Arthritis

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A patient with chronic rheumatoid arthritis was admitted to the hospital with cervical fracture. Two weeks after acute steroid withdrawal, she was noted to have pleural effusion, and signs and symptoms of acute pericardial tamponade. An open partial pericardiectomy and drainage relieved the symptoms. The sequence of events suggests that rapid steroid withdrawal might have precipitated rapid accumulation of pleural and pericardial fluid.

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The spectrum of rheumatoid pericardial involvement includes subclinical pericarditis, clinical fibrinous pericarditis, pericarditis with effusion and constrictive pericarditis. Cardiac tamponade secondary to rheumatoid arthritis is a relatively rare entity. A patient with classical rheumatoid arthritis who developed acute cardiac tamponade two weeks after abruptly discontinuing steroids is presented.

Case Report

A 60-year-old woman was admitted to the orthopedic service on January 15, 1976 after an automobile rear end collision in which she sustained "whiplash" injury. The clinical examination and x-ray studies revealed no significant bony or soft tissue injury except a fracture of the odentoid process of the axis without displacement. She gave a five-year history of rheumatoid arthritis and the features of the disease included morning stiffness, pain on motion, and deformities of multiple joints, swelling of the proximal metacarpo-phalangeal, elbow and knee joints bilaterally. Her symptoms had been controlled by the use of salicylates and prednisone, 15 mg daily.

On admission to the hospital, the patient had a "halo cast" applied for the fracture of the cervical vertebrae, and steroids were discontinued. Her initial chest x-ray film revealed normal cardiac size and configuration with normal lung fields (Fig 1). The electrocardiogram was unremarkable. The patient's sedimentation rate was 60 mm (Westergren), hematocrit 30 percent, and the red cells were hypochromic. The BUN, glucose, and electrolytes were within normal limits. The RA latex test in the serum was positive in 1:640 dilution. The antinuclear antibodies and the lupus erythematosus preparations were negative, and the platelet count was within normal limits. The third component of the complement fixation test was 87 mg (normal 80-140 mg). The SGOT was 42 units, the LDH 160 units, and the CPK 44

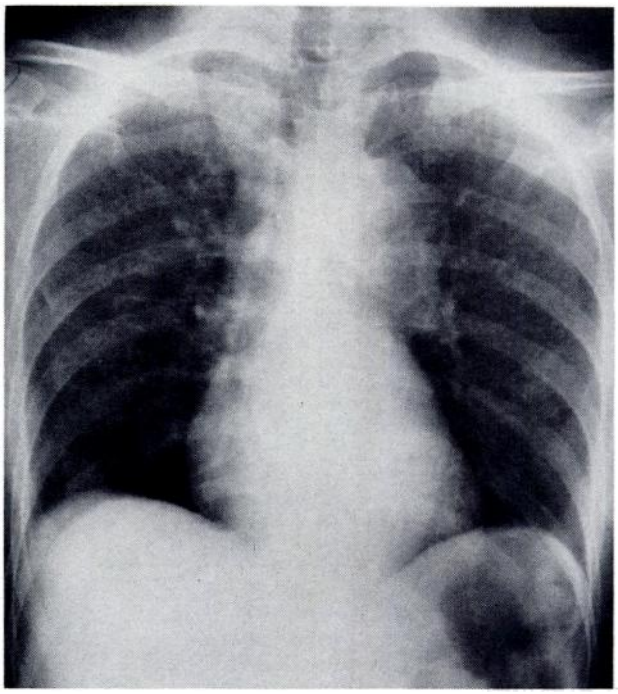

Figure 1. The chest x-ray film on admission. Cardiac size and configuration are within normal limits.