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Chest 1970;57;333-335
DOI 10.1378/chest.57.4.333

ISSN:0012-3692

Cystic Medionecrosis of the Pulmonary Arteries*

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Cystic medionecrosis (CMN) characteristically affects the aorta. That it also may involve vessels in the lung is not widely appreciated. Two patients are presented in whom cystic medionecrosis of the pulmonary arteries accounted for dramatic clinical and pathologic findings. One had massive dilatation of both main pulmonary arteries together with a healed dissecting aneurysm extending from the aortic arch to the iliac bifurcation. The other had an enormous saccular aneurysm of the left main pulmonary artery.

Cystic medionecrosis (CMN) is a specific type of degeneration involving elastic fibers and muscle elements within the media of the arterial wall.[1,2] It is characterized histopathologically by numerous mucoid cysts filled with metachromatically staining material.[3] The affected artery is predisposed to aneurysmal dilatation, medial dissection, and spontaneous rupture.[4-6]

Cystic medionecrosis characteristically occurs in the aortae of patients with the Marfan syndrome.[3,4,7] Sometimes, however, it appears as an isolated arterial lesion.[8-10]

This communication concerns two patients in whom CMN of the pulmonary arteries caused striking clinicopathologic findings.

Case Reports

Case 1

A 63-year-old Negro woman entered the hospital because of increasing pedal edema and breathlessness of three weeks' duration. Additional historical information never became available.

The patient appeared critically ill. Her blood pressure was 150/70 mm Hg; pulse rate, 70 beats per minute; respiratory rate, 40 per minute; and oral temperature, 99°F. She weighed 155 pounds and her body habitus was normal. She had no specific skeletal or ocular manifestations of the Marfan syndrome. Her jugular venous pressure was greatly elevated. The precordium was hyperdynamic with an apical impulse in the midaxillary line. A grade III/VI systolic ejection murmur and a IV/VI diastolic decrescendo murmur were loudest to the left of the sternum in the third and fourth interspaces. Rales were audible at the bases of both lungs. A nontender hepatic edge was palpable 10 cm below the right costal margin. Both legs were massively edematous. Carotid, brachial, and femoral arterial pulsations were normal, bilaterally.

An electrocardiogram showed atrial fibrillation, occasional ventricular premature contractions, and a pattern of right ventricular hypertrophy. Chest roentgenogram (Fig 1) disclosed cardiomegaly, pulmonary venous congestion, and large, bilateral hilar masses.

On the second hospital day catheterization of the right side of the heart revealed a pulmonary arterial pressure of 98/35 mm Hg (mean, 60 mm Hg) and a simultaneous brachial arterial pressure of 142/59 mm Hg (mean, 89 mm Hg). Attempts to measure pulmonary capillary wedge pressure were unsuccessful. Angiography (Fig 2) demonstrated massive dilatation of the major pulmonary arteries; visualization of the more distal branches was poor.

The patient's status rapidly worsened. Multiple pulmonary thromboemboli seemed likely. Three days after admission, thoracotomy with pulmonary arteriotomy revealed no thromboemboli. Shortly thereafter, the patient suffered cardiac arrest and died.

Autopsy disclosed aneurysmal dilatation of the main pulmonary arteries and a healed dissecting aneurysm extending from the arch of the aorta to the iliac bifurcation. Cystic medionecrosis was evident throughout the pulmonary arterial tree as well as the aorta and its major branches. Cause of death was not apparent.

Case 2

A 19-year-old Negro man was admitted to the hospital complaining of shortness of breath and easy fatiguing of eight years' duration. He had suffered from recurrent upper respiratory infections throughout childhood.

On physical examination the patient appeared much younger than his chronological age. He was 62 inches tall and weighed 80 pounds. His pulse rate was 70 beats per minute; respiratory rate, 19 per minute; blood pressure, 100/70 mm Hg; and oral temperature, 99°F. He had a webbed neck and an asymmetrical chest with prominence

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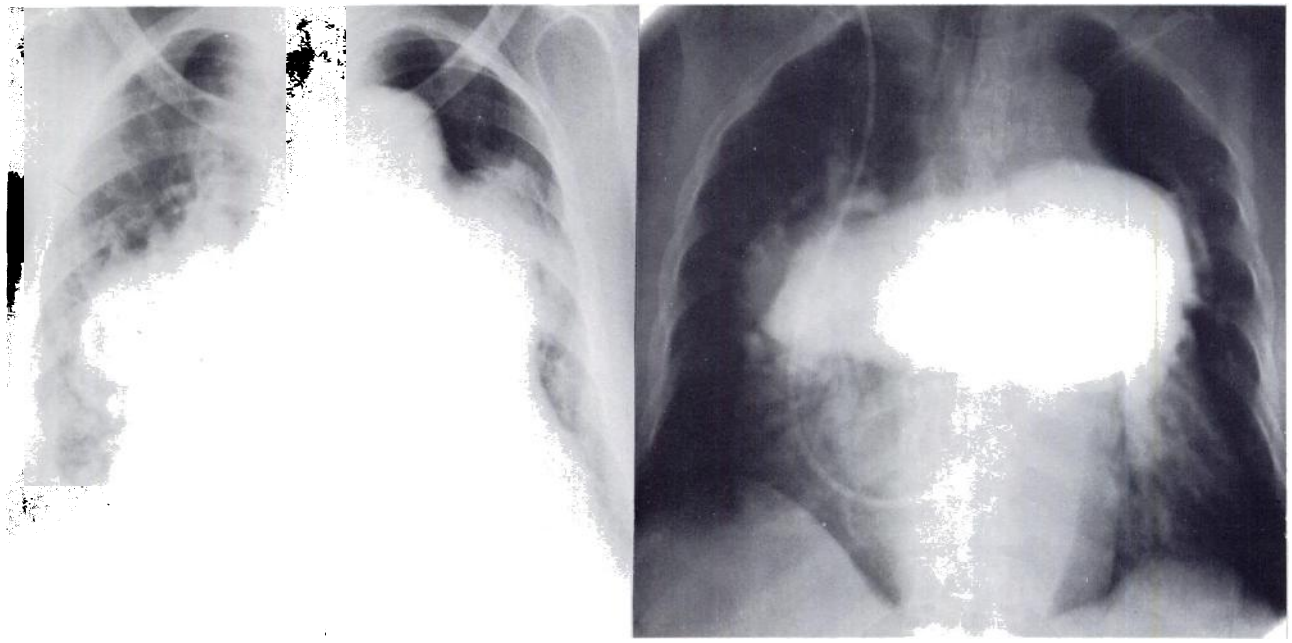

FIGURE 1 (Case 1), left. Chest roentgenogram depicting cardiomegaly and bilateral hilar masses. FIGURE 2 (Case 1), right. Angiogram demonstrating enormously dilated major pulmonary arteries.

of the left hemithorax. The precordium was active with a right and left ventricular lift. A low pitched to-and-fro murmur was audible in the fourth left intercostal space.

Chest roentgenogram (Fig 3) demonstrated dilatation of the right pulmonary artery and a large density filling the anterior mediastinum and much of the left hemithorax. Electrocardiogram showed a sinus rhythm and a pattern of biventricular hypertrophy with right atrial enlargement. Cardiac catheterization revealed a pulmonary arterial pressure of 50/25 mm Hg (mean, 40 mm Hg) and a simultaneous aortic pressure of 95/70 mm Hg. Angiocardiogram demonstrated a left-to-right shunt through a ventricular septal defect; the catheter delineated a large, saccular aneurysm of the left main pulmonary artery (Fig 4).

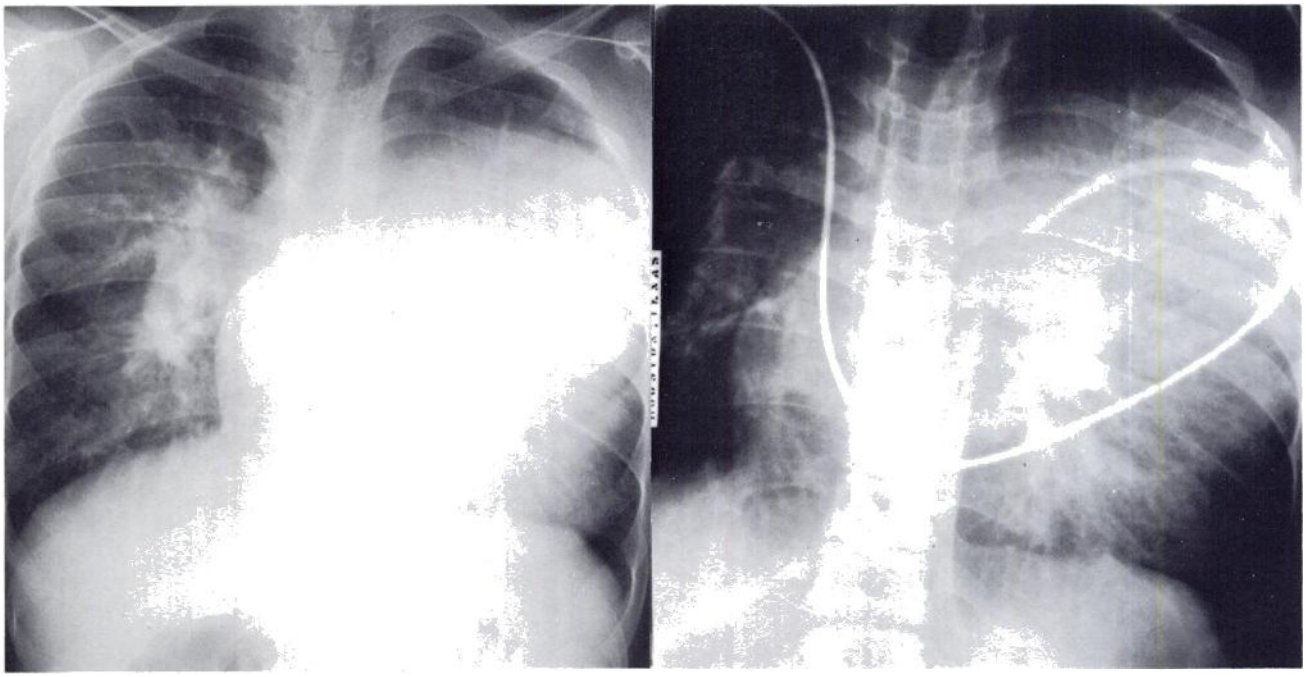

FIGURE 3 (Case 2). Chest roentgenogram showing dilatation of right pulmonary artery and large mass in left hemithorax. FIGURE 4 (Case 2). Chest roentgenogram disclosing angiocatheter coursing through right atrium and right ventricle before entering main pulmonary artery. Essentially all of catheter to left of spine lies within large saccular aneurysm of left main pulmonary artery.

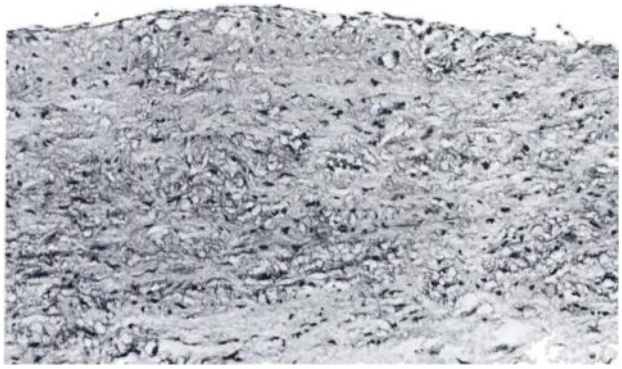

FIGURE 5 (Case 2). Photomicrograph of pulmonary artery demonstrating extensive microcystic change of the media. (Hematoxylin and eosin, original magnification × 360.)

At thoracotomy an interventricular septal defect 1.5 cm in diameter was closed. The aneurysm of the left main pulmonary artery was repaired by partial resection and angioplasty; its wall contained changes typical of cystic medionecrosis (Fig 5).

COMMENTS

These two patients are noteworthy because pulmonary arterial disease incident to CMN dominated their clinical presentation. Moreover, successful repair of a pulmonary arterial aneurysm resulting from CMN has not been documented previously.

Detailed information on CMN involving the pulmonary vasculature is sparse. In the Marfan syndrome the spectrum of such lesions ranges from idiopathic dilatation of the pulmonary arteries[11] and minimal abnormalities of the elastic fibers of the media[3,12-14] to typical CMN.[4-8] Only rarely do such changes overshadow those in the aorta.[15-17] Cystic medionecrosis of the pulmonary arteries also occurs in association with long-standing pulmonary hypertension[6] and leads occasionally to dissection and rupture of the affected vessels.[6,16,18] The relation between pulmonary hypertension and CMN of the pulmonary vasculature remains unclear.

Widely disseminated cystic medionecrosis in our first patient and phenotypic resemblance of the second patient to others with unusual manifestations of the Marfan syndrome[10] suggest congenital rather than acquired arterial defects. We recognize, however, that the vascular lesions in both patients may have had no connection with the Marfan syndrome.

ACKNOWLEDGMENT: Drs. J. F. Howell, S. A. Kinard, J. O. F. Roehm, Jr., and R. E. Fechner permitted us to present the second patient.

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