A Case of Marked Dilatation of the Pulmonary Arterial Tree Associated with Mitral Stenosis

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A case which was studied with the technic of catheterization is discussed as a problem in the diagnosis of this syndrome.

A CASE of marked dilatation of the entire pulmonary arterial tree with calcification is being reported because of the rarity of the condition, and the unusual roentgenologic and clinical findings. The patient has been followed at the Michael Reese Hospital for a number of years. Intracardiac catheterization was recently performed in an attempt to arrive at a more accurate diagnosis, and to determine, if possible, what primary etiologic factors were involved.

Case Report

Mrs. A. C., a 42 year old housewife, was admitted to the Michael Reese Hospital on November 27, 1948, with minor lacerations and bruising following an automobile accident. Her father had died of pneumonia, and her mother of a heart attack at the age of 42 years. As a child her development was normal, and she enjoyed fairly good health. At the age of 14 years her tonsils were removed. Signs of heart disease present at that time led to a diagnosis of mitral stenosis. At the age of 16 she developed acute rheumatic fever and spent eight months in bed. It was during this illness that cyanosis and clubbing of the fingers were first noted. After this episode, however, aside from slight dyspnea on exertion, she carried on fairly normally, and continued to work. She was married at the age of 24. During the next few years she had four miscarriages.

In March 1939 she spent six weeks in the hospital with pneumonia, and developed thrombophlebitis of the veins of the leg. Physical examination of the cardiovascular system at that time revealed a diffuse apical impulse in the sixth intercostal space at the anterior axillary line. The second pulmonic sound was accentuated. There was a systolic thrill and a harsh systolic murmur over the left border of the sternum, and a systolic and presystolic murmur at the apex. The liver and spleen were not palpable. There was slight ankle edema, cyanosis was present, and there was marked clubbing of the fingers.

In November 1939 a pregnancy was terminated because of the patient's cardiac lesion. One month later, following a cold and sore throat, she was readmitted to the hospital with fever, polyarthritis, dyspnea, orthopnea, and cyanosis. She developed acute pericarditis with effusion and congestive failure. Her course was stormy requiring oxygen therapy, mercurial diuretics, and digitalization. After discharge from the hospital, dyspnea on walking persisted and ankle edema was now a frequent manifestation. In February 1943 she was again admitted to the hospital, this time for medical examination before undergoing extraction of a tooth. At that time her main complaints were frequent headaches, mild dyspnea on exertion, and occasional ankle edema. She had been taking digitalis steadily for three years. Physical examination revealed a well nourished, well developed cyanotic woman. There was marked clubbing of the fingers and toes. A malar flush was present. Her blood pressure was 140/90. A few moist rales were present at both lung bases. There was a systolic retraction of the lower end of the sternum. The apex beat was diffuse. The heart was enlarged to the left and downward to the sixth intercostal space just outside the midelavicular line. A systolic thrill was palpable in the third left intercostal space, lateral to the sternum border. The second pulmonic sound was very loud. There was a short, harsh systolic murmur at the apex with accentuation of the first sound. This was followed by a low-pitched, rumbling mid-diastolic murmur. A rough systolic murmur of a different quality was present over the left border of the sternum, best heard over the lower portion. The liver border was palpable 5 cm. below the right costal margin and pulsation was present. The spleen was not palpable.

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MARKED DILATATION OF PULMONARY ARTERIAL TREE

There was slight pitting edema about the ankles. Fluoroscopy revealed a normal-sized left ventricle. Both auricles were markedly enlarged. Both inflow and outflow tracts of the right ventricle were enlarged. Marked dilatation of the pulmonary artery, affecting the trunk and both main branches, with calcification in the walls, was noted. Laboratory studies revealed a hemoglobin value of 14.3 grams with 6,260,000 red blood cells. The blood Kahn reaction was negative. Treatment consisted of bed rest, diuretics, and digitalis.

In June 1945 the patient was again hospitalized because of severe congestive failure, with a history of cold, cough, and sore throat for six weeks prior to admission. While in the hospital, she developed thrombophlebitis of the legs. Soon after, several episodes of acute chest pain followed by hemoptysis occurred and bilateral ligation of the femoral veins was performed. In November 1946 she was again hospitalized because of an exacerbation of congestive heart failure. In 1947 she was hospitalized on two occasions because of sudden chest pain, cough, and blood-streaked sputum, and a diagnosis of pulmonary emboli was entertained. Two other admissions in 1947, and three in 1948 were mainly for recurrent upper respiratory infections and exacerbation of heart failure. However, repeated pulmonary embolization may have played an important role during these episodes, which were marked by frequent attacks of sudden chest pain and hemoptysis.

Physical examination, made on admission in November 1948 following the automobile accident, revealed multiple bruises, cuts, and ecchymoses of the face, chest, and limbs. The patient was orthopneic and cyanosed. The point of maximum impulse was in the sixth intercostal space at the anterior axillary line. Findings on auscultation were similar to those heard on previous admissions. A phonocardiogram, recorded with the Sanborn Stetho-Cardiette, showed a systolic murmur after the first sound at the pulmonic and aortic areas. Systolic and mid-diastolic murmurs were recorded at the apex. The veins of the neck were distended. Moist rales were present at the lung bases. The liver was enlarged and tender, extending to 7 cm. below the right costal margin. The spleen was tender and palpable with deep inspiration. There was moderate ankle and sacral edema. The earliest available electrocardiogram, made in 1939 when she had pneumonia, showed a right-sided heart strain. A second electrocardiogram, made in February 1940 during convalescence from a severe episode of acute rheumatic fever, pancreatitis, and congestive failure, showed the development of a block of the right bundle branch system. An electrocardiogram made during her last admission showed no significant changes from previous tracings. Fluoroscopic findings were similar to those of previous admissions.

A review of this patient’s chest films revealed that marked aneurysmal dilatation of the pulmonary artery trunk and its two major branches was present as far back as 1933. The chest films exposed in 1940 showed a dense biloebular mass the size of a lime with incomplete calcification of the periphery in the right midlung field. The main trunk and left branch of the pulmonary artery were diffusely dilated. The radiologist who then had access to films taken in 1933 and 1936, noted that little change in the size of the lesions had occurred. In the earlier films, however, there had been no evidence of

Fig. 1.—Teleoroentgenogram of the chest made in July 1948. Posteroanterior (A, Left) and left anterior oblique, (B, Right) views. (Discussed in text.)
calcification. Roentgenograms made during the patient's illness in 1943 showed an increase in size of the mass in the right midlung field, with less tendency to lobulation, and an increase in calcium deposit. No appreciable radiologic changes occurred in the next five years and roentgenograms made in 1948 (fig. 1) revealed no changes, showing chiefly three dense shadows within the chest, one in the right midlung field and two in the left side of the chest closer to the hilus.

This patient was seen by a number of different observers during her numerous admissions to the hospital. The most likely diagnosis considered was aneurysmal dilatation of the pulmonary artery and mitral stenosis. The possibility of an associated interauricular septal defect (Lutembacher's syndrome) was strongly entertained. The masses did not pulsate, however, and even with the aid of fluoroscopy, one could not rule out the possibility of lung cyst or tumor. The possibility of an A-V aneurysm was remote.

**Physiologic Studies.** It was decided that intracardiac catheterization would be of diagnostic help in this case, especially in determining the presence or absence of interauricular septal defect or other possible congenital shunt. The technic of rightsided heart catheterization as described by Courmand and colleagues was closely followed. Blood samples were withdrawn in quick succession from the right and main pulmonary arteries, the right ventricle, right auricle, and superior vena cava. Roentgenograms were made and pressures recorded with a Hamilton manometer at each position of the catheter. At the conclusion of catheterization, a sample of blood was withdrawn from the femoral artery. The oxygen content of the blood samples was determined by the method of van Slyke and Neill. Blood-flow estimations were based on the principle outlined.

![Fig. 2.—Anteroposterior films exposed with the patient in the supine position at a distance of four feet, by the Potter-Bucky technic. A (Left), The catheter has passed along the markedly dilated right pulmonary artery. B (Right), The tip of the catheter in the right ventricle demonstrates the tremendous size of that chamber.](image)

**Table 1.—Summary of Data**

<table>
<thead>
<tr>
<th>Catheter Location</th>
<th>Oxygen Vols. %</th>
<th>Systolic Pressure (Mm. Hg)</th>
<th>Diastolic Pressure (Mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right pulm. art.</td>
<td>9.5</td>
<td>80</td>
<td>35</td>
</tr>
<tr>
<td>Main pulm. art.</td>
<td>10.9</td>
<td>80</td>
<td>35</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>Sample</td>
<td>75</td>
<td>12</td>
</tr>
<tr>
<td>Right auricle</td>
<td>9.5</td>
<td>mean pressure</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>10.1</td>
<td>10.6</td>
<td>+5</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>12.9</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Resting oxygen consumption = 200 cc. per minute.

Cardiac output = \[
\frac{200}{12.9 - 10.1} \times 100 = 6.9 \text{ liters per minute.}
\]

by Fick. The patient's oxygen consumption was determined with a Sanborn Metabolator.

Results of catheterization are shown in table 1. The oxygen content of blood samples from the superior vena cava, the right auricle, and the pulmonary artery did not differ significantly. Pressures were definitely elevated in the pulmonary artery and right ventricle. The oxygen content of arterial
blood drawn from the femoral artery was 12.9 volumes per cent. The oxygen capacity was 19.4 volumes per cent and the arterial oxygen saturation was calculated to be 66.5 per cent. The patient's resting oxygen consumption was 200 c.c. per minute. By dividing the oxygen consumption per minute by the arteriovenous oxygen difference of the blood in the right auricle and the femoral artery the cardiac output was estimated to be 0.9 liters per minute. Figure 2, A shows the catheter in the markedly dilated right pulmonary artery and identifies it beyond doubt. In figure 2, B the tip of the catheter is in the right ventricle (as confirmed by pressure curves and blood oxygen content) and demonstrates the tremendous size of that chamber.

The catheterization studies led to the following conclusions: (1) Roentgenologic visualization of the catheter in the large masses established their identity as parts of a dilated pulmonary arterial tree. (2) Since the blood samples taken from the superior vena cava, the right auricle, and the pulmonary artery did not differ significantly in oxygen content, the possibility of the presence of a patent ductus arteriosus or of a septal defect was ruled out. (3) The marked unsaturation of the peripheral arterial blood strongly suggested the presence of pulmonary arteriolar disease with poor respiratory gas exchange. The relatively high cardiac output in all probability acted as a compensatory mechanism, similarly to what is noted in chronic cor pulmonale and in emphysema. (4) The high pressures in the pulmonary artery and right ventricle were likely due to a combination of (a) mechanical obstruction at the mitral valve, and (b) increased peripheral pulmonary resistance by an altered pulmonary arteriolar bed. The equality of systolic pressure in the pulmonary artery and right ventricle eliminated the possibility of pulmonary stenosis.

COMMENTS

Clinical and physiologic studies would indicate that the main lesions present in this case were: (1) Marked aneurysmal dilatation of the main pulmonary artery and its major branches with calcification of the arterial walls. (2) Associated rheumatic heart disease with mitral stenosis. (3) An altered pulmonary arteriolar bed which may be primary, or secondary, to the mitral stenosis.

Greene and his co-workers have recently reported 4 patients with idiopathic congenital dilatation of the pulmonary artery, apparently the first group of subjects in which physiologic studies were performed with the aid of intracardiac catheterization. They found normal pressures in the right ventricle and lower pressures in the pulmonary artery. They attributed the lower pulmonary arterial pressure to turbulence created by the deformity, to an increase in expansibility of the pulmonary arteries due to the thinness and dilatation of their walls, or to a relative stenosis caused by a stretching of the free edges of the semilunar cusps across the orifice of the valve. As mentioned above, in the patient here reported, the pressures obtained in the pulmonary artery are grossly elevated, therefore pointing to the presence of associated lesions.

The association of dilatation of the pulmonary artery and mitral stenosis has been described before. D'Aunoy and von Haam, in their review of 85 patients with pulmonary aneurysms, noted 5 cases that were associated with mitral stenosis. In all 5, the pulmonary artery trunk alone was involved. In 4 of the 5 cases, sclerosis of the pulmonary artery, of unknown origin, was considered to be the main etiologic factor. The fifth case was one of congenital mitral stenosis, and a congenital defect in the pulmonary vasculature was believed responsible for the aneurysm.

Atheromatosis of the pulmonary arterial tree as well as pulmonary arteriolar sclerosis has been noted fairly frequently in association with dilatation of the pulmonary artery. Deterling and Clagett, in a review of 36 cases of aneurysm of the pulmonary artery proved by necropsy, noted the presence of atheromas in 11 of the patients. In 3 out of these 11, there was marked arteriosclerosis throughout the lungs associated with right cardiac dilatation. They reported a patient of their own with aneurysmal dilatation of the right pulmonary artery, and severe sclerosis of the arterioles in both lung fields. Gibson described a patient with a huge pulmonary artery, who after death was shown to have a primary proliferative arteriolar sclerosis of the pulmonary vessels. Gold reported a case of congenital dilatation of the pulmonary artery trunk and the branches, associated with arteriosclerotic changes throughout the entire pulmonary arterial tree. On the other hand, Parker and Weiss, in reporting on the structural changes in the lungs in mitral stenosis, described intimal proliferative changes in the larger
branches of the pulmonary arterial tree, marked fibrous thickening of the intima of the medium-sized arteries, and a hyperplastic arteriosclerosis of the arterioles.

On the basis of pathologic criteria set up by previous workers, one is not justified in regarding the present case as one of pure congenital dilatation of the pulmonary artery. On the other hand, it seems extremely unlikely that mitral stenosis, no matter how severe, even when giving rise to pulmonary arteriolar sclerosis and marked pulmonary hypertension, could, per se, lead to such massive diffuse dilatation of the pulmonary arteries. It is therefore believed that an inherent weakness, present in the arterial wall since birth, was a primary factor in allowing the dilatation to occur. Whether the pulmonary arteriolar changes which were suggested by this study were secondary to the mitral stenosis, or were congenital in origin remains unknown.

SUMMARY

1. A case of marked diffuse dilatation of the pulmonary arterial tree associated with calcification in the walls has been presented.
2. The difficulties in determining the primary etiologic factors have been discussed.
3. The results of intracardiac catheterization have been described, and the conclusions reached have been discussed.

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REFERENCES

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