Clinical Studies on Involvement of the Pulmonary Artery by Syphilitic Aortic Aneurysms

By John J. Donnell, M.D., David C. Levinson, M.D. and George C. Griffith, M.D.

Cardiac catheterization and angiocardiography studies are reported in two patients with syphilitic aortic aneurysms, both of which compromised the pulmonary circulation. In the first instance the right main pulmonary artery was compressed and pulmonary hypertension proximal to the compression resulted. In the second patient the aneurysm ruptured into the pulmonary artery producing an aortic-pulmonary fistula. This patient also had pulmonary hypertension and evidence at cardiac catheterization of a large left-to-right shunt. The difficulty of making the clinical diagnosis of pulmonary artery compression by an aortic aneurysm is discussed. Cardiac catheterization and angiocardiography were essential for establishing this diagnosis in life.

The purpose of this report is to describe two cases of syphilitic aortic aneurysm, each of which compromised the pulmonary artery circulation. In one case, the aneurysm created chronic cor pulmonale by obstructing the pulmonary artery with extrinsic pressure; in the other case, the aneurysm ruptured into the pulmonary artery, producing a chronic aortic-pulmonary fistula. Despite the frequency of syphilitic aortic aneurysms, the above complications have been rarely diagnosed during life.

Usually, involvement of the pulmonary circulation becomes apparent at the time that the aneurysm ruptures into the pulmonary artery. Clinically, this event is acute and disastrous as a rule, but occasionally the rupture may occur with survival and present signs suggesting patent ductus arteriosus.1-3 Nicholson4 in 1943, reviewed the literature, consisting of 81 reported cases, and added two cases of rupture of syphilitic aortic aneurysm into the pulmonary artery. Subsequently, additional cases have been reported.4-6 More than likely, most cases of this type are preceded by pulmonary artery compression.3 In 1939, Garwin and Siegal8 reported three cases of cor pulmonale due to compression of the pulmonary artery without rupture of the aneurysm. In these instances, death resulted from what appeared to be right sided heart failure. Later, Eichler and Heller9 and Pearson and Nichol10 reported similar cases. Under any circumstance, the diagnosis is a difficult one, as indicated by the latter authors. Abrahams and Wood11 reported a case thought preoperatively to have pulmonary stenosis, who had positive serology as well. At surgery, an unidentified obstruction was found within the pulmonary artery along with hypertension proximal to the obstruction. An aortic aneurysm was not identified.

The following cases are thought to be of interest because the diagnoses were made clinically and verified by both cardiac catheterization and angiocardiography.

Case Report

Case 1: The patient was a 38-year-old Negro man, admitted to the Los Angeles County Hospital on Nov. 7, 1951 because of left anterior chest pain of six months' duration. The pain had become more severe during the previous two months, and he had observed aggravation by coughing and sneezing. Some relief was afforded by resting in the supine position. He had noted shortness of breath only when the pain was especially severe. There was a history of penile chancre at the age of 17, which healed spontaneously. Incidental discovery of positive serology at the age of 32 resulted in “a few” injections, thought to be penicillin. At the age of 25 he was injured in an automobile accident, which he thought resulted in fractures of “several ribs” on the right side.

The patient was a well developed, well nourished Negro, complaining of chest pain, but was not
acutely ill. The blood pressure was 140/70 mm. Hg in the right arm and 135/56 in the left arm. Radial pulses were equal and normal in volume. The pupils were normal, and eye grounds were negative. A systolic pulsation in the suprasternal notch was noted. A tracheal tug was not present. An area of systolic expansile pulsation 2 by 3 cm. in size could be seen in the second left intercostal space about 3 cm. from the sternal border, and a systolic thrill was palpable at this point. The point of maximum impulse was in the fifth intercostal space at the midclavicular line; the left border of cardiac dullness extended about 1 cm. lateral to this point. Both heart sounds were heard at all valve areas, and the second pulmonic was louder than the second aortic sound. A grade III systolic murmur was described at the pulmonic area, and a faint diminuendo diastolic murmur was heard along the left sternal border. The heart rate was 80 beats per minute, with a regular rhythm interrupted occasionally by premature contractions. The lungs were clear. The examination of the abdomen was negative. A penile scar was noted. No dependent edema was present, and peripheral pulsations were unimpaired. No neurological abnormality was demonstrable.

Hemoglobin was 14 Gm. per 100 cc. There were 9800 white blood cells per cubic millimeter. Urinalysis was negative. The Wassermann was strongly positive. Examination of spinal fluid gave negative serology and colloidal gold; protein was 38 mg. per 100 cc.

Roentgenogram of chest showed an aneurysm of the ascending aorta and arch (fig. 1). The electrocardiogram showed right axis deviation in a vertical type heart.

**Cardiac Catheterization:** Under local anesthesia a No. 8 French cardiac catheter was introduced into the left basilic vein in the median antecubital space and passed successively under fluoroscopic guidance into the superior vena cava, right atrium, right ventricle, main pulmonary artery and right pulmonary artery. An abrupt rise in pressure was noted as the catheter was being withdrawn from the right pulmonary artery toward the main pulmonary artery. This maneuver was repeated several times, and the same pressure change occurred constantly (fig. 2).

Data obtained by cardiac catheterization have been tabulated in table 1. The findings at cardiac catheterization established moderate right ventricular hypertension, hypertension of the main pulmonary artery, and "acquired stenosis" of the right pulmonary artery, with reduced pressure distal to compression by aneurysm.

**Angiocardiogram:** With the patient in the supine position, 50 cc. of 70 per cent Neo-iopax was rapidly injected into the left basilic vein and six x-ray exposures obtained at two-second intervals. The dye entered the superior vena cava, right atrium, right ventricle and the main pulmonary artery. The dye passed into the left pulmonary artery, but the right pulmonary artery did not fill (fig. 3). The aneurysm and the left ventricle were well visualized (fig. 4). Angiocardiography confirmed that a large aneurysm of the aortic arch was obstructing the main right pulmonary artery.

**Follow-up:** On March 28, 1952, the patient was readmitted for the purpose of wiring the aneurysm. By the cutaneous route, 100 feet of No. 32 wire were inserted into the aneurysmal sac. One week later, 200 feet more were inserted, and the patient was discharged subsequently.

On Feb. 18, 1953, he returned to the hospital because of a peri rectal abscess. This required incision and drainage, and was later excised. At that time, he had no complaints which were related to his aneurysm, and his physical findings were essentially
TABLE 1.—Findings at Cardiac Catheterization
Case 1

<table>
<thead>
<tr>
<th>Station</th>
<th>B.P. (mm.Hg)</th>
<th>Oxygen Vol. (%)</th>
<th>Saturat. (%)</th>
<th>Additional Data</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior Vena Cava</td>
<td>3/1</td>
<td>10.4</td>
<td>59.3</td>
<td>O₂ Cap. 17.6 vol.%</td>
</tr>
<tr>
<td>Right Atrium (Mid)</td>
<td>3/1</td>
<td>10.1</td>
<td>57.6</td>
<td>O₂ Cons. 180 cc./min.</td>
</tr>
<tr>
<td>Right Atrium (High)</td>
<td>3/1</td>
<td>10.7</td>
<td>60.6</td>
<td>Cardiac Output 3.6 L./min.</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>50/7</td>
<td>10.2</td>
<td>58.0</td>
<td></td>
</tr>
<tr>
<td>Main Pulmonary Artery</td>
<td>50/12</td>
<td>9.6</td>
<td>54.6</td>
<td></td>
</tr>
<tr>
<td>Right Pulmonary Artery (Proximal to stenosis)</td>
<td>40/14</td>
<td>10.1</td>
<td>57.5</td>
<td></td>
</tr>
<tr>
<td>Right Pulmonary Artery (Distal to stenosis)</td>
<td>15/8</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Femoral Artery</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

FIG. 3

FIG. 4

FIG. 3. (Case 1): Posterior-anterior angiocardiogram showing dye in right atrium, right ventricle, main pulmonary artery and left pulmonary artery with nonvisualization of the right pulmonary artery.

FIG. 4. (Case 1): Posterior-anterior angiocardiogram showing visualization of the left ventricle and aortic aneurysm 12 seconds after injection of the dye.

unchanged. The systolic thrill and murmur were described, but no diastolic murmur was found. The blood pressure was recorded at 120/70. The chest x-ray and electrocardiogram were unchanged.

Case 2: A 46-year-old Negro woman entered the hospital on April 13, 1954, with the chief complaints of dyspnea, orthopnea and swelling of the legs and abdomen. Onset of her present illness occurred in August, 1953, at which time she developed paroxysmal episodes of dyspnea with wheezing, associated with "sickness" in the upper abdomen. These attacks were brief in duration, sudden in onset and cessation, and seemed to be initiated by assuming the left lateral decubitus position. There was no relation of their onset to effort. In between these episodes she felt well, except for persistent anorexia with resultant weight loss. In November, 1953, she was forced to discontinue her occupation as a domestic. Swelling of the ankles and exertional dyspnea first were noted in Jan., 1954, progressing to dyspnea at rest, orthopnea and marked swelling of the legs and abdomen by April, 1954. There was no history of a heart murmur, cyanosis, rheumatic fever or syphilis.

The patient was a normally developed, but emaciated, orthopneic Negress who was severely dyspneic,
and preferred to lie on her right side. Her voice was quite hoarse. The pupils were round, equal and normally reactive to light and accommodation. The neck veins were distended in the upright position. The trachea was displaced slightly to the left, and a prominent tracheal tug was noted. A systolic lift of the upper sternum was apparent, unassociated with localized pulsation or a well identified point of maximum impulse. A systolic thrill, felt maximally at the third and fourth left intercostal spaces, was transmitted widely over the precordium. Impaired resonance was found at the manubrium, along with a widening in the upper mediastinum on percussion. The left border of cardiac dullness extended about 9 cm. to the left of the mid-sternal line in the fifth intercostal space. Valve sounds were normal at all areas, except for splitting and accentuation of the pulmonic second sound. A continuous murmur with systolic accentuation, best heard at the fourth left intercostal space next to the sternum, was transmitted widely over the chest, but especially into the left infraclavicular area and axilla. The systolic component was grade IV to V in intensity. The cardiac rhythm was regular at a rate of 120 beats per minute. The blood pressure was 180/80-0 mm. Hg in each arm, and the pulse was Corrigan (water-hammer) in quality. Decreased resonance was found in the right lung base, along with bilateral basal crepitant rales. Sonorous rales and wheezes were confined to the left posterior chest, and there was accentuation of these sounds during cardiac systole; this phenomenon was accentuated when the patient was tilted toward the left, during which maneuver a paroxysm of coughing occurred. Free fluid was demonstrable in the abdomen, and a tender liver was palpable four cm. below the right costal margin.

Hemoglobin was 11 Gm. per 100 cc. There were 7400 white blood cells per cubic millimeter. Urinalysis was negative. The Wassermann reaction was strongly positive.

Roentgenogram of chest showed grade I left ventricular enlargement and aneurysm of the arch and first portion of the descending thoracic aorta (fig. 5). An electrocardiogram was suggestive of early left ventricular hypertrophy.

Response to specific cardiotherapy was satisfactory. With restoration of cardiac compensation, 27 pounds of edema fluid were eliminated. Dyspnea and orthopnea disappeared, but the patient continued to prefer to lie on the right side. The blood pressure spontaneously reduced to 118/50-30 mm. Hg.

Cardiac Catheterization: Under local anesthesia a No. 7 French cardiac catheter was introduced into the thoracic aorta and the aorta passed under fluoroscopic guidance successively through the superior vena cava, right atrium, right ventricle, main pulmonary artery, and finally into the right peripheral pulmonary artery. Pressures and blood samples were taken at these stations. An attempt was made to insert the catheter into the left lung field, but it could not be passed beyond the left main pulmonary artery. Data obtained by cardiac catheterization have been tabulated in table 2. The findings at cardiac catheterization established (1) definite evidence of a left to right shunt into the pulmonary artery and (2) elevation of pressures in the right ventricle, pulmonary artery, and wedged peripheral pulmonary artery, possibly secondary to left ventricular failure.

Angiocardiograms: Two studies were carried out, the first in the anterior-posterior projection and the second in the left oblique. In both studies, 50 cc of 70 per cent Neo-iopax was rapidly injected into the left basilic vein. In the anterior-posterior projection films were exposed at the rate of one per second. Neo-iopax passed readily into the right pulmonary artery, but the left pulmonary artery was compressed by a large mass (fig. 6). In the left oblique view exposures were at the rate of two per second. Radiopaque material was visible in the main pulmonary artery and in branches of the right pulmonary artery (fig. 7). Dye was not visualized in the left lung field. Also a dilated and elongated arch was apparent with aneurysmal dilatation of the first portion of the descending aorta. At approximately 12 seconds after the dye was injected, it was visualized in the arch of the aorta (fig. 8).

These studies confirmed the diagnosis of a chronic aortic-pulmonary fistula, resulting from the rupture of a syphilitic aortic aneurysm into the pulmonary
Table 2.—Findings at Cardiac Catheterization

**Case 2**

<table>
<thead>
<tr>
<th>Station</th>
<th>Pressure (mm.Hg)</th>
<th>Oxygen Vol.%</th>
<th>Oxygen Satur.%</th>
<th>Additional Data</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior Vena Cava</td>
<td>8/0</td>
<td>8.63</td>
<td>30.59</td>
<td></td>
</tr>
<tr>
<td>Right Atrium (Mid) (Low)</td>
<td>8/0</td>
<td>7.85</td>
<td>46.01</td>
<td></td>
</tr>
<tr>
<td>Right Ventricle (Body) (Conus)</td>
<td>85/7</td>
<td>8.93</td>
<td>52.35</td>
<td></td>
</tr>
<tr>
<td>Main Pulmonary Artery</td>
<td>80/35</td>
<td>8.39</td>
<td>49.18</td>
<td>O₂ Cap. 17.06 vol.%</td>
</tr>
<tr>
<td>Right Pulmonary Artery</td>
<td>75/32</td>
<td>7.85</td>
<td>46.01</td>
<td>O₂ Cons. 125 cc./min.</td>
</tr>
<tr>
<td>Left Pulmonary Artery</td>
<td>85/32</td>
<td>13.24</td>
<td>77.61</td>
<td>Systemic blood flow 1.7 L./min.</td>
</tr>
<tr>
<td>Right Peripheral Pulmonary Artery</td>
<td>25/20</td>
<td>13.45</td>
<td>78.84</td>
<td>Shunt flow 6.5 L./min.</td>
</tr>
<tr>
<td>Right Femoral Artery</td>
<td></td>
<td>15.29</td>
<td>89.62</td>
<td>Cardiac Output 8.2 L./min.</td>
</tr>
</tbody>
</table>

**Fig. 6.** (Case 2): Posterior and anterior angiogram showing dye in the main pulmonary artery and compression of the left pulmonary artery by the aneurysm.

**Fig. 7.** (Case 2): Posterior-anterior angiogram in the left oblique view showing dye in the main pulmonary artery and in branches of the right pulmonary artery with non-visualization of the branches of the left pulmonary artery.

artery. Exploratory thoracotomy was performed on May 6, 1954. The following is a report of the surgery and findings.

Under general anesthesia, the patient was placed on the right side and a parascapular incision was made in the skin. Dissection was carried down through the subcutaneous tissue and chest muscle to expose the ribs. The fifth rib was resected subperiosteally and pleural space entered. Mediastinal pleura overlying the aneurysm and pulmonary artery was opened and dissection carried out around the aorta and pulmonary artery to a point where any further resection would have resulted in a massive bleeding. There was marked fusiform dilatation of the ascending and transverse arch. The beginning descending aorta was about 4 to 5 times normal in size. A marked thrill was felt over the pulmonary artery which could be obliterated by closing off the left pulmonary artery. The pulmonary artery was also markedly dilated and there was marked inflammatory reaction between aorta and pulmonary artery,
defect, made the diagnosis seem obvious. Although rupture of an aortic aneurysm into either of the heart chambers or great vessels may produce this same murmur and thrill, individual features characterizing the various sites of rupture exist. Herrmann and Schofield have emphasized these individual features, and have delineated the specific syndrome of rupture of an aortic root-aneurysm into the right atrium. Cardiac catheterization produced conclusive evidence of a left to right shunt into the pulmonary artery in this case, and angiocardio- gram revealed compression with displacement of the left main pulmonary artery. These findings were also confirmed by a thoracotomy.

Summary

Two cases of syphilitic aortic aneurysm are presented, each involved the pulmonary artery. Cardiac catheterization and angiography were essential in establishing the specific abnormalities present in each case. These two techniques are well suited to the study and clarification of such problems. We are unable to find any previous reports wherein such an opportunity presented itself. As the prospect for surgical correction of such complications improves, the need for accurate diagnosis increases.

Summario in Interlingua

Es reportate studios de catheterisation cardiaca e de angiocardiographia in duo patientes con syphilitic aneurysmas aortic compromitente le circulation pulmonar. In le prime caso le major arteria dextero-pulmonar esseva comprimite con le resultato de hypertension pulmonar in le segmento cis-compressional. In le secunde caso le aneurysma se rumpeva a in le arteria pulmonar con le resultato de un fistula aortico-pulmonar. In iste caso hypertension pulmonar esseva etiam constatale, e le catheterisation cardiac revelava signos del presentia de un considerabile derivation sinistro-dextere. Es discutite le difficultate de establir le diagnose de compression pulmono-arterial per aneurysma aortic. Nos trovarque que catheterisation cardiac e angiocardiographia esseva indispen-
sabile pro establir iste diagnose durante le vita del paciente.

REFERENCES

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