Direct Communication of a Pulmonary Artery with the Left Atrium
An Unusual Variant of Pulmonary Arteriovenous Fistula

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The clinical interest evidenced in recent years in pulmonary arteriovenous fistula results in part from the availability of curative surgical procedures for its treatment. Since over 140 cases of this condition have been reported in recent years, it can no longer be considered rare. In pulmonary arteriovenous fistula a direct communication exists between a large pulmonary artery, on the one hand, and a pulmonary vein on the other. Therefore, pulmonary arterial blood low in oxygen content is delivered without further oxygenation through the fistula to the left atrium. The result of this right-to-left shunt may be cyanosis, clubbing, dyspnea, polycythemia, and the opportunity for paradoxical embolization to the systemic circuit. The brain seems particularly prone to receive such emboli. Pulmonary arteriovenous fistulas range in size from very small, without clinical findings, to very large communications with prominent clinical manifestations. This report presents the case of a patient with an unusual form of pulmonary arteriovenous fistula, namely, direct communication of the right lower pulmonary artery with the left atrium. This anomaly had evidently been responsible for a large right-to-left shunt, as cyanosis had been a prominent clinical sign in this patient who succumbed to a cerebral abscess.

Case Report

Clinical Findings

A 3-year-old, cyanotic white female child awoke one evening complaining that her left arm hurt. Shortly thereafter, the arm began to shake and in 1 hour she was unable to move it. Subsequently, several mild convulsive movements of the left arm and left leg occurred. The past history revealed that a normal delivery had followed a normal pregnancy. Growth and development had been normal but the patient was noted to be "dusky" at 1 year of age. No other symptoms were referable to the cardiopulmonary system.

Physical examination revealed a cyanotic child who was listless, but could be aroused. The body temperature was 104 F. by rectum. Clubbing of the fingers and toes was present. A soft, systolic murmur was heard over the base of the heart. The cardiac rate and rhythm and blood pressures were within normal limits. The only abnormal neurologic findings were a left knee jerk more active than the right and absent abdominal reflexes.

Laboratory studies revealed a hemoglobin of 18.5 Gm. per 100 ml., a hematocrit level of 62 per cent, and a total leukocyte count of 8,600 per cu. mm. The differential leukocyte count in percentages was as follows: neutrophils, 71; lymphocytes, 25; monocytes, 3; and eosinophils, 1. The sedimentation rate was 15 mm. per hour. No remarkable findings were observed in the urinalysis, blood chemical or spinal fluid studies. Cultures of the urine and of the throat revealed no significant features. A Mantoux tuberculin test gave negative results. An electroencephalogram suggested a localized lesion in the right cerebral hemisphere.

An electrocardiogram (fig. 1) revealed left axis deviation and evidences of left ventricular hypertrophy and left atrial enlargement. A thoracic roentgenogram (fig. 2) suggested slight generalized cardiac enlargement with prominence of the left atrium and minimal right ventricular enlargement. A rounded density approximately 3 em. in diame-
Electrocardiogram taken during initial period of hospitalization. The pertinent findings include left axis deviation of the QRS complex and evidences of left ventricular hypertrophy and left atrial enlargement.

The left ventricle, which could not be separated from the cardiac shadow, was present just to the right of, and anterior to, the spine. The pulmonary vascular markings were interpreted as normal.

The admitting diagnosis was congenital cardiac disease with right-to-left shunt and cerebral abscess. Penicillin and chloramphenicol were administered. On the second day of hospitalization the temperature had fallen below 100 degrees and remained normal thereafter. The neurologic symptoms cleared during the following 12 days. Results of repeated blood counts and urinalysis were normal throughout 12 days of antibiotic therapy. A thrombotic or embolic episode of the brain was therefore considered more likely than an infectious one. The antibiotics were discontinued. On discharge from the hospital, on the fourteenth day, the patient's neurologic findings were normal. She was to return in 1 month for angiocardiographic confirmation and treatment of a clinically suspected pulmonary arteriovenous fistula.

Three weeks following discharge from the hospital the patient suddenly became febrile and comatose. The spinal fluid contained 70,000 polymorphonuclear leukocytes per cubic millimeter. Despite intensive antibiotic and supportive treatment, the patient died 6 hours following the onset of the terminal illness.

Pathologic Findings

The pertinent findings were restricted to the central nervous and cardiovascular systems. Meningitis was associated with a solitary abscess in the right cerebral hemisphere containing 60 ml of purulent material. Culture of this material revealed Pseudomonas aeruginosa.

Study of the cardiovascular system revealed that the pulmonary trunk and the aorta were normally interrelated. The wall of the pulmonary trunk was thick, being almost as thick as the wall of the aorta.

The origins of the right upper and lower pulmonary arteries and of the left pulmonary artery each showed a zone of moderate stenosis beyond which the walls approached normalcy in thickness (fig. 3, left).

The left lung and the course of the left pulmonary artery were normal. The left pulmonary veins, two in number, followed a normal course to the left atrium. The right lung had only one lobe, the upper, which filled the right hemithorax. The one vein from this lobe followed the course of the right upper pulmonary vein and entered the left atrium normally.

The right pulmonary artery bifurcated into upper and lower branches. The upper branch entered the single lobe of the right lung. The right lower pulmonary artery led into the left upper aspect of a thin-walled sac-like structure.

Left. Posteroanterior thoracic roentgenogram taken during initial period of hospitalization. The pulmonary vascular markings are within normal limits. The bulge along the left cardiac border suggests left atrial enlargement. A rounded density is seen to the right of the spine in the region of the right pulmonary hilus. This density could not be separated from the cardiac shadow but was thought to represent a pulmonary arteriovenous fistula. Right. Lateral roentgenogram. The rounded density in question lies behind and inferior to the position of the left atrium.
measuring about 2.5 by 2 cm. in diameter. This structure, which lay to the right of the left atrium, was imbedded in a concavity of the medial aspect of the adjacent enlarged single right pulmonary lobe. The lower medial aspect of the sac-like structure, which received the right lower pulmonary artery, communicated with the left atrium at the expected location of the ostium of the right lower pulmonary vein (fig. 3, right). Because of the latter connection, on the one hand, and the aforementioned connection with the right lower pulmonary artery, on the other, the sac-like structure provided an unbroken connection between the right lower pulmonary artery and the left atrium (fig. 4).

The right ventricular wall was of normal thickness, whereas the left ventricular wall was considered mildly hypertrophied as its wall measured about 1.4 cm. in thickness. The left atrial and ventricular cavities did not appear enlarged.

Discussion

As in the case reported, communication between a pulmonary artery and the left atrium is to be considered a form of pulmonary arteriovenous fistula. In the usual form of this condition, pulmonary parenchyma is formed, and the abnormal communication lies within pulmonary tissue. In our case, absence of the middle and lower lobes in association with the fistula described suggests that the primordia of these lobes had at one time been present but that while the vascular connections persisted, the parenchyma failed to develop. Enlargement of the capillary bed of the anomalous area of the right lung may be represented by the sac-like structure described. The arterial connection with this structure is
representative of the early arterial system in this area, while the connection with the left atrium may be looked upon as representing the right lower pulmonary vein.

Since direct communication of the right lower pulmonary artery with the left atrium appears to represent a form of pulmonary arteriovenous fistula, a brief consideration of this larger category of congenital disease is in order. The recent excellent reviews on pulmonary arteriovenous fistula by Stringer and associates and by Muri have well summarized the cases reported in the literature. Both reviews found that about 50 per cent of cases of pulmonary arteriovenous fistula are associated with telangiectasis of the skin and mucous membranes. Cyanosis and clubbing of the digits were very frequent. Dyspnea was less common, and was manifested relatively late. Congestive heart failure was unusual. Neurologic symptoms were frequent, occurring in 41 of the 140 cases reviewed by Stringer, including five patients who succumbed with cerebral abscess. A systolic cardiac murmur was present in about half the patients. Hemoglobin and hematocrit values were commonly elevated. Despite frequent and pronounced cyanosis, cardiomegaly either by roentgenographic or by electrocardiographic criteria was unusual.

An insight into the natural history of the anomaly may be gained from Muri’s review of 117 cases of pulmonary arteriovenous fistula. Of the 50 patients who did not have corrective surgery, 20 succumbed, including five with cerebral abscess and seven with rupture of the fistula. Primary surgical mortality in the 67 patients who had received surgical correction was 7 per cent.

The diagnosis of a pulmonary arteriovenous fistula may frequently be made from the thoracic roentgenogram by visualization of a mass either near the pulmonary hilus or in a peripheral pulmonary field. This diagnosis may be confirmed by venous angiocardiography, which demonstrates the shunt from the pulmonary arterial to the pulmonary venous systems.

Friedlich and associates reported on the hemodynamic findings in four patients with pulmonary arteriovenous fistulas (one patient with direct communication between the right pulmonary artery and the left atrium). Normal pulmonary arterial pressure was universal. Peripheral arterial oxygen desaturation resulted from a right-to-left shunt which was calculated to vary from 42 to 76 per cent of the right ventricular output. Despite this large diversion of blood, cardiac output was normal in three patients and only slightly above normal in the fourth.

Estimates of resistance of the fistulous tract alone and of the pulmonary vascular bed, exclusive of the fistula, were made by the aforementioned investigators. Resistance in the fistula was equal to that seen in the normal pulmonary vascular bed, whereas the vascular resistance in the nonfistulous part of the pulmonary vascular system was about twice normal. Since the normal and anomalous pulmonary circuits are in parallel, this difference of resistance favors flow through the low re-
sistance (anomalous) channel. The common phenomenon of delay in appearance of cyanosis until adolescence or adult life is perhaps on a basis of acquired increasing resistance in the nonfistulous part of the pulmonary vascular bed.

The type of pulmonary arteriovenous fistula wherein the pulmonary artery communicates directly with the left atrium is a rare abnormality. Only two other cases are known to the authors. One of these occurring in a 15-year-old boy was reported by Friedlich and associates\(^3\) as well as by Sloan and associates.\(^4\) In this patient, cyanosis was first apparent at 5 years of age and gradually increased in intensity. Clubbing of the digits and polycythemia were associated. A pulsating hemangioma was present on the forehead. Cardiac catheterization revealed a normal right ventricular output but it was estimated that 71 per cent of this output was shunted into the systemic circulation (via the left atrium). The electrocardiogram showed right axis deviation, and the thoracic roentgenogram revealed a normal cardiac shadow with no abnormalities of the pulmonary fields. Angiocardiography demonstrated a right-to-left shunt in close proximity to the right pulmonary hilus.

At operation performed on this patient by Dr. Alfred Blalock, it was reported that one of the branches of the right pulmonary artery, which measured about 1 cm. in diameter, extended posteriorly and communicated directly with the left atrium. When the anomalous vessel was interrupted, the patient’s cyanosis disappeared immediately.

A second case of communication of a pulmonary artery with the left atrium was reported in the Case Records of the Massachusetts General Hospital.\(^5\) A 45-year-old woman was admitted with mental confusion. The history revealed that she had been cyanotic for at least 7 years. Examination revealed generalized cyanosis but no clubbing of the digits. Congestive heart failure and atrial fibrillation were apparent. The electrocardiogram revealed evidence of right ventricular hypertrophy and right bundle-branch block and evidence for anterior myocardial infarction. Supportive treatment failed and the patient succumbed. On pathologic examination there was a direct communication of the right pulmonary artery to the upper portion of the left atrium, the circumference of the anomalous channel being 2.5 cm. In addition, there were cerebral emboli and infarcts, emboli in the posterior descending coronary artery, and splenic infarcts.

The clinical findings in patients with direct communication of a pulmonary artery with the left atrium differ only in degree from those in the more common types of pulmonary arteriovenous fistula. The large diameter of the fistulous channel in this type of connection and its short course results in a low resistance in the fistulous channel and facilitates a large right-to-left shunt.

The cerebral abscess present in our case and its common occurrence in intrapulmonary types of arteriovenous fistula is considered a complication of the circumstances of a central right-to-left shunt. In regard to this particular complication, pulmonary arteriovenous fistula is similar to the many forms of intracardiac malformations that are associated with a right-to-left shunt.\(^6\), \(^7\)

The electrocardiogram of the patient whose case is here reported is of interest. It revealed evidence of left atrial enlargement, left axis deviation, and left ventricular hypertrophy. While the electrocardiogram has not been stressed in other reports on pulmonary arteriovenous fistulas, it has usually been described as normal. In our case, the findings of left atrial and left ventricular hypertrophy may, in fact, be due to the large right-to-left shunt that had apparently been present with a resultant augmentation of blood flow through the left atrium and left ventricle.

It is apparent from the study of the pathologic material in our case that surgical ablation of the fistulous channel at an appropriate time would have resulted in elimination of the right-to-left shunt and would have prevented the fatal complication, the cerebral abscess.
Summary

Direct communication of a pulmonary artery with the left atrium is described and considered a variant of pulmonary arteriovenous fistula.

A 3-year-old girl presented with clinical evidence of a lesion of the central nervous system. The history revealed that she had been cyanotic and had had clubbing of the digits and polycythemia. A rounded mass was noted in the region of the right pulmonary hilus in the posteroanterior thoracic roentgenogram. A pulmonary arteriovenous fistula was suspected, but the patient succumbed with a cerebral abscess before definitive diagnostic studies and therapy were undertaken.

Anatomic examination revealed an unusual variant of pulmonary arteriovenous fistula, namely, direct communication of the right lower pulmonary artery with the left atrium and absence of the middle and lower lobes of the right lung.

It is important that pulmonary arteriovenous fistulas be suspected clinically, since definitive diagnosis and therapy are now possible. While the hemodynamic effects of this type of lesion are usually not severe, untreated patients suffer a high morbidity and mortality through systemic arterial oxygen desaturation, paradoxical embolization, cerebral abscess, and rupture of the fistula.

References

From the materialistic and the energetic standpoint alike, carbon, hydrogen, and oxygen, each by itself, and all taken together, possess unique and preeminent chemical fitness for the organic mechanism. They alone are best fitted to form it and to set it in motion; and their stable compounds, water and carbonic acid, which make up the changeless environment, protect and renew it, forever drawing fresh energy from the sunshine.

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