Vascular Malformation Overlying the Pulmonary Artery Simulating a Patent Ductus Arteriosus

By Calvin B. Ernst, M.D., Karl P. Klassen, M.D., and Joseph M. Ryan, M.D.

The presence of an asymptomatic patent ductus arteriosus is usually suspected on the basis of auscultatory findings, and the diagnosis is supported by electrocardiography and roentgenography. Other vascular and cardiac lesions that produce continuous murmurs to the left of the upper sternum are usually symptomatic and are associated with additional changes requiring cardiac catheterization and contrast studies for a final diagnosis. Aneurysmal dilatation of the coronary vessels associated with arteriovenous fistulas give rise to murmurs simulating patent ductus arteriosus. These anomalies may be entirely asymptomatic or cause the symptoms of myocardial ischemia. The following is a case report of a coronary-pulmonary artery fistula not involving the myocardium and simulating a patent ductus arteriosus.

Case Report

S. S., hospital no. 691383. An 18-year-old college girl was referred for evaluation of a heart murmur first heard 6 months previously during a routine examination. Growth and development since birth had been normal and the patient had been asymptomatic.

On physical examination the abnormal physical findings were limited to the heart. The temperature was normal with a pulse of 80 and a respiratory rate of 20 per minute. The blood pressure in the right arm was 110/70 mm. of mercury, and in the left arm 150/72 mm. The heart was not enlarged. The point of maximum cardiac impulse was in the fifth intercostal space 1 cm. medial to the left midclavicular line, and a thrill was not felt. A continuous grade III systolic, early grade II diastolic, crescendo-decrescendo murmur was heard loudest in the second intercostal space 2 cm. to the left of the sternum. The murmur was well localized and was not transmitted to the axilla, back, or precordium; it was rather soft and could be heard best with the patient leaning forward. There was no clubbing or cyanosis.

Laboratory determinations were all within normal limits. Cardiac fluoroscopy showed the heart to be normal in size and general configuration. The electrocardiogram showed only slight right axis deviation.

The consensus was that this represented a small patent ductus arteriosus. It was thought that thoracotomy was indicated because of the location and type of the murmur. Through a left postero-lateral thoracotomy, the mediastinum was exposed in the region of the ductus arteriosus, and the thrill was located more centrally along the pulmonary artery. Pressure on the mediastinum in the region of the ductus did not obliterate the thrill, whereas pressure on the proximal pulmonary artery did. The mediastinum was opened and a fibrous ligamentum arteriosum was found; it was transected and found not to be patent.

When the pericardium was opened posterior to the phrenic nerve, multiple serpiginous dilated vessels were seen over the base of the pulmonary artery extending to the point where the pulmonary artery leaves the pericardium (fig. 1). Aneurysmal dilatation was present in several areas on the pulmonary artery where a thrill was felt. The vascular anomaly did not involve the myocardium. With gentle compression of the aneurysm, the thrill disappeared. The tortuous vessels were fed by three arterial branches, two of these coming from the right and left coronary arteries, and one from the region of the pericardiophrenic artery as the pulmonary artery leaves the pericardium. There were other small branches from accessory vessels from the left atrium and the conus artery. Temporary occlusion of the feeder vessels produced no observable color changes in the myocardium and no abnormalities in the electrocardiographic tracing of the monitor.

A small incision was made in one of the aneurysmal dilatations, and bright red blood spurted at systolic pressure. The feeders were then ligated with collapse of the aneurysms and disappearance of the thrill. The patient did well postoperatively and was discharged seven days later without the heart murmur.

Discussion

Excellent discussions concerning the rela-
Appearance of coronary-pulmonary artery fistula prior to and subsequent to multiple ligation.

Relatively rare congenital coronary arteriovenous fistulas have appeared in the literature. The majority of the reported cases represent arteriovenous fistulas with flow from the involved coronary arteries to the right atrium, right ventricle, or pulmonary artery, thus representing a left-to-right shunt. The diagnosis was based on autopsy findings prior to 1947, when Björck reported the first such case diagnosed and treated by ligation during a thoracotomy performed for a suspected patent ductus arteriosus. Since that time the diagnosis of coronary arteriovenous fistula has been made at the time of thoracotomy. Several cases have been reported in which a correct diagnosis was made on the basis of cardiac catheterization and angiography.

The present case differs from previously reported ones in that the arteriovenous malformation did not involve the walls of the ventricle or atrium but only the main pulmonary artery. Blood flow was in the direction of the pulmonary artery, as proved by the presence of high pressure oxygenated blood in the supplying branches. Both coronary vessels, which were normal in distribution and size, supplied branches to the fistula and there was an additional vessel coming from the pericardiophrenic artery or possibly from branches of the bronchial artery along the pulmonary artery.

It has been established that the walls of the pulmonary artery and veins within the pericardium derive their blood supply from branches of the coronary vessels while the extrapericardial portion of the pulmonary artery and veins receive their blood supply from the bronchial arteries. These arterial and venous vasa vasorum do not communicate with the lumen of the pulmonary artery under normal conditions as do vasa vasorum of the systemic arteries. In pulmonic stenosis in tetralogy of Fallot or following thrombosis of the pulmonary vessels, perforation of the wall of the pulmonary vessels by these vasa vasorum can and probably does occur. In the present case, however, we were unable to determine the reason for such fistula formation; there was no evidence of pulmonary stenosis or of previous thrombosis. The small size of the aneurysm and the supplying vessels explained the absence of symptoms and the normal cardiac size and pulmonary vascularity. Catheterization of this patient might have indicated the presence of this small shunt; retrograde aortography of good quality, however, would have led to a correct preoperative diagnosis.

Summary

An arteriovenous fistula with aneurysmal dilatation involving branches of both coronary vessels and the pulmonary artery is discussed.

The malformation was limited to the main pulmonary artery and presented a small left-to-right shunt without symptoms.

Ligation of the supplying vessels resulted in the disappearance of a murmur typical of a patent ductus arteriosus.
MALFORMATION SIMULATING PATENT DUCTUS

References

Constancy of the External Environment

The regulatory devices of our modern laboratories have not yet succeeded in rivaling the ocean. Singly, certain conditions, for example, temperature, alkalinity, and concentration, may be more accurately regulated by man, though on a small scale only; but the regulation of all such properties together is not yet possible. The only known improvement upon the ocean is the body of a higher warm-blooded animal. Here, however, the processes of organic evolution have begun with the ocean, and in several respects merely perfected existing arrangements.—Lawrence J. Henderson. The Fitness of the Environment. New York, The MacMillan Co., 1924, p. 186.
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