Anomalous left coronary artery from the main pulmonary trunk: physiologic and clinical importance of its association with patent ductus arteriosus and pulmonary hypertension

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Origin of the left main coronary artery from the pulmonary trunk is an extremely rare, fatal, but potentially treatable anomaly. Left ventricular perfusion with desaturated blood with low pressure from the pulmonary artery results in early death. Associated pulmonary hypertension can help to prevent ischemia. We present a four-month-old boy with this anomaly associated with patent ductus arteriosus and pulmonary hypertension.

Key words: anomalous left coronary artery, pulmonary hypertension.

Origin of the left coronary artery (LCA) from the pulmonary trunk (Bland-White-Garland syndrome) is an extremely rare but important anomaly because early death is often the natural outcome. In this report, we present a four-month-old case with this entity hat had been protected from early neonatal death by associated patent ductus arteriosus (PDA) and pulmonary hypertension.

Case Report

A four-month-old boy was admitted to our hospital with the chief complaints of cyanosis upon crying, dyspnea and seizures. The neonatal period had been uneventful and these symptoms had begun after the age of three months. The history did not reveal any abnormalities of feeding, easy fatigability or excessive sweating with minor effort.

The child appeared well nourished, with length and weight measurements over 90th percentile for age. Physical examination was normal except for mild peripheral cyanosis. Complete blood count and blood gas analysis were normal. The chest X-ray showed moderate cardiomegaly with increased pulmonary vascularity. Electrocardiography showed normal sinus rhythm, marked right axis deviation, right ventricular hypertrophy, ST segment depression in precordial leads V_{4R} and V_1, and absent Q waves in left precordial leads V_4-V_6.

On echocardiography, the right heart chambers and main pulmonary artery were extremely enlarged and a second-degree tricuspid regurgitation with a peak velocity of 4.6 m/sec was observed, which led to the diagnosis of pulmonary hypertension. Fractional shortening was 0.39; all other aspects of the examination were normal. Echocardiographic evaluation of the coronary ostia was not performed, and the patient was subjected to cardiac catheterization to identify the cause of pulmonary hypertension.

On catheterisation, the pulmonary arterial pressure was 71/42, mean 56 mmHg; aortic pressure was 97/37, mean 58 mmHg; and the left atrial pressure was not elevated. The pulmonary artery O_2 saturation was 77%. Ductal patency had been demonstrated by the passage of the catheter retrogradely from the aorta. The ascending aortogram did not show the patent ductus arteriosus which might have been because of the evident pulmonary hypertension. The origin and course of the right coronary artery was as normal in the aortic root injection, but the left coronary artery was not seen. A main pulmonary artery angiogram demonstrated the anomalous left coronary artery from the

pulmonary trunk with anterograde flow into the anomalous left coronary artery from the main pulmonary artery (Fig. 1). With these findings, a diagnosis of Bland-White-Garland syndrome with patent ductus arteriosus and pulmonary hypertension was made. The patient was scheduled for dissection and reimplantation of the left main coronary artery to the appropriate sinus of Valsalva when he was lost suddenly just before surgery.

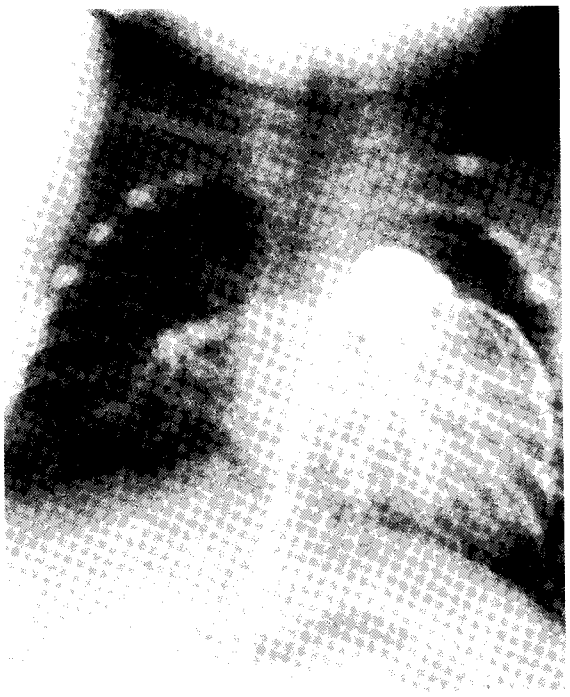

Fig. 1. Angiographic view of the left main coronary artery originating from the pulmonary trunk.

Discussion

Anomalous origin of the LCA from the pulmonary trunk is an extremely rare anomaly, having been found in only 24 out of 23,249 coronary angiograms done in older children and adults[1]. Of all children born with this anomaly, about 87% present in infancy and about 65 to 85% die before one year of age from intractable congestive heart failure, usually after two months of age[2].

This anomaly is well tolerated in fetal life, because pressures and oxygen saturations are similar in the aorta and the pulmonary artery. After birth, the pulmonary artery contains desaturated blood at a rapidly falling pressure; accordinly, the left ventricle is perfused with desaturated blood at low pressures. At first, ischemia is transient and occurs only with exertion, such as feeding or crying, but further increase in myocardial oxygen demand leads to infarction of the anterolateral left ventricular free wall. This causes congestive heart failure, which is often made worse by a dilated mitral ring or infarction of the anterior papillary muscle. Myocardial ischemia stimulates the development of collateral circulation between the two coronary artery systems. The severity of the symptoms and survival beyond this point will depend on the adequacy of this collateral circulation. In about 15% of these patients, myocardial perfusion can sustain myocardial function at rest or even during exercise and these cases can reach adult life.

This anomaly is often isolated, but has been associated with PDA, ventricular septal defect, tetralogy of Fallot, and coarctation of the aorta. The presence of an associated congenital heart disease may either mask the clinical picture of the anomalous LCA with its own symptoms or provide higher pressure perfusion to the coronary circulation via the pulmonary artery, such as in the presence of a PDA and pulmonary hypertension or pulmonary hypertension alone. A large arterial duct will maintain adequate left coronary perfusion and thus delay presentation until the duct is ligated. Indeed in some cases it could be a cause of sudden deterioration or cardiac arrest[3,4].

Pulmonary hypertension not associated with congenital heart malformations, pulmonary parenchymal disease, left atrial hypertension, hypoventilation, or other known causes of pulmonary hypertension, are encountered rarely in adults, and even less frequently in infants and children. Newborns with increased pulmonary vascular resistance and right-to-left shunting are usually considered to have persistent pulmonary hypertension of the newborn as in our case, whereas older infants and children are said to have "primary" or "unexplained" pulmonary hypertension[5].

On physical examination, there may or may not be evidence of congestive heart failure. Radiologically, there is marked cardiomegaly and pulmonary edema. Thallium-901 myocardial perfusion imaging shows reduced uptake in the anterolateral ischemic region. Because the patient usually presents with an anterolateral

infarct, there may be abnormal Q waves in leads I, aVL, and V_4-V_6. There may also be abnormal R waves or R wave progression in the left precordial leads. However, these findings are not specific, because they are encountered in cardiomyopathies or in myocardial infarcts from other causes as well. Our case showed, instead, right axis deviation, right ventricular hypertrophy, and right ventricular strain which are the electrocardiographic (ECG) findings of associated pulmonary hypertension.

Echocardiography is replacing cardiac catheterisation as the standard method of diagnosis. Abnormal attachment of the LCA can be seen, and the direction of coronary arterial blood flow toward the great artery establishes the diagnosis. Enlarged right coronary artery, size and function of the cardiac chambers, regional left ventricular wall motion abnormalities, mitral regurgitation and echogenicity of te papillary muscles and adjacent endocardium can be demonstrated as well. Cardiac catheterisation and angiography are employed only if echocardiographic results are uncertain[6]. However, we emphasized the importance of evaluating the coronary ostia in every patient on echocardiographic examination in order not to miss a potentially treatable anomaly.

This anomaly can be treated with effective surgery. The principal surgical methods are ligation of the origin of LCA to prevent pulmonary-coronary steal, ligation of the origin of LCA with reconstitution of flow with a subclavian arterial or saphenous venous graft, direct reimplantation of the origin of LCA to the aorta, or creation of an aortopulmonary window and a tunnel that directs blood from the aorta to the left coronary ostium. The late results after surgery are fairly good. The heart becomes smaller, congestive heart failure abates, left ventricular shortening fraction improves, and mitral incompetence tends to regress[7].

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