Congenital Aneurysm of the Left Sinus of Valsalva With an Aortopulmonary Tunnel

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Aneurysm of the left sinus of Valsalva is rare, and there is only one previous report of rupture into the pulmonary artery. This report describes a patient with valvular pulmonary atresia and ventricular septal defect in whom a portion of his pulmonary blood flow was supplied by an aortopulmonary tunnel arising from a left sinus of Valsalva aneurysm. The surgical implications of precise definition of the type of aortopulmonary communication are discussed.

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Congenital aneurysms of the sinus of Valsalva usually involve the noncoronary sinus, less frequently the right coronary sinus and rarely the left coronary sinus (1). Rupture of a sinus of Valsalva aneurysm generally results in a fistulous connection between the aorta and a contiguous structure, most often the right atrium or right ventricle. An aortoatrial or aortoventricular tunnel may result from organization of fistulous connections created by rupture of the sinus of Valsalva aneurysm occurring in utero (2). We report an unusual case with a left sinus of Valsalva aneurysm and an aortopulmonary tunnel.

Case Report

History and physical examination. The patient is an 8 year old white boy first seen at our hospital at 2 years of age. He had undergone cardiac catheterization at another hospital at 3 months of age and a diagnosis of pulmonary atresia with ventricular septal defect was made. A communication between the aorta and the main pulmonary artery was also found and was considered to be a left coronary artery to pulmonary artery fistula. He was asymptomatic with normal growth and development. There was mild cyanosis and clubbing.

Cardiac examination showed a prominent right ventricular impulse, normal first heart sound and a loud and single second heart sound. A grade 3/6 harsh pansystolic murmur and a grade 2/6 continuous murmur were heard best at the mid-left sternal border and upper left sternal border, respectively. Continuous bruits were present over both lung fields. Liver and spleen were not palpable. Peripheral pulses were bounding.

Cardiac catheterization. Repeat cardiac catheterization at 8 years of age showed a small left to right atrial shunt, a bidirectional ventricular shunt and large left to right aortopulmonary shunts from both the ascending and descending aorta. Aortic saturation was 90%. Right ventricular pressure was at systemic level. The pulmonary artery was not entered, but bilateral pulmonary venous wedge pressures were normal. Cineangiography showed valvular pulmonary atresia, large subaortic ventricular septal defect, overriding aorta, small atrial septal defect and left aortic arch. A typical windsock appearance of a large left sinus of Valsalva aneurysm was demonstrated (Fig. 1). The pulmonary blood supply was given by four large bronchial collateral arteries as well as by a fistulous connection between the aneurysm and the small main pulmonary artery (Fig. 2). Coronary angiography showed a normal left coronary artery (Fig. 3) that arose from the sinus of Valsalva aneurysm, 2.9 cm from the aortic root.

Other studies. Cross-sectional images of the great arteries obtained by two-dimensional echocardiography (Fig. 4) and ultrafast computed tomography (Fig. 5) also demonstrated the left sinus of Valsalva aneurysm and the aortopulmonary tunnel.

Current status. The patient remains asymptomatic and has not yet undergone surgery.

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Discussion

Differential diagnosis. The differential diagnosis of nonsurgical communications between the ascending aorta and main pulmonary artery includes truncus arteriosus and aortopulmonary window, both of which are direct connections. Indirect connections include anomalous origin of the left or right coronary artery from the pulmonary artery, coronary artery fistula, rupture of sinus of Valsalva aneurysm into the pulmonary artery and aortopulmonary tunnel.

There are two previous reports of coronary artery fistula to the pulmonary artery (3,4), both in patients with pulmonary atresia and ventricular septal defect. In each, the sinus of Valsalva was normal and the connecting fistula arose from a coronary artery. The first report of rupture of a sinus of Valsalva aneurysm into the pulmonary artery in an adult with an otherwise normal heart appeared recently (5). The aneurysm involved the left sinus of Valsalva; the coronary artery anatomy was not reported. The present case is the first report of a left sinus of Valsalva aneurysm and aortopulmonary tunnel. The normal coronary anatomy ruled out a coronary artery fistula.

Figure 1. Angiogram with contrast injection into the right ventricle (RV), frontal projection. The aorta (Ao) opacified from a large right to left shunt through the ventricular septal defect. The left sinus of Valsalva aneurysm (An) was to the left of the aortic root. L = left pulmonary artery; R = right pulmonary artery.

Figure 2. Angiograms with contrast injection into the left sinus of Valsalva aneurysm (An). A, Lateral projection, B, corresponding line drawing and C, frontal projection with cranial tilt. The aneurysm was posterior (A,B) and to the left (C) of the small main pulmonary artery (MPA). The aortopulmonary tunnel (T) is best seen in A. The left coronary artery (LCA) also opacified from the aneurysm. CA = coronary artery; L = left pulmonary artery; R = right pulmonary artery.
Surgical implications. The distinction between an aortopulmonary tunnel arising from a sinus of Valsalva aneurysm and a coronary artery fistula communicating with the pulmonary artery has important surgical implications. Rastelli et al. (4), described a coronary artery fistula that could be ligated with impunity. In the present case there is a high level of flow through the aneurysm and tunnel into the pulmonary artery. Ligation of the tunnel without repair of the aneurysm might result in sluggish flow in the aneurysm, thrombus formation and, possibly, severe compromise of left coronary artery flow. The potential for rupture of the unrepaired aneurysm must also be considered. Transaortic repair of the aneurysm would require considerable mobilization of the left coronary artery. If that proved impossible, the surgeon might need to reroute flow to the left coronary artery through a graft. The long-term prognosis of coronary bypass grafts in children is unknown.

This case is, to our knowledge, the first of its type reported. It illustrates the importance of careful delineation of the nature of an aortopulmonary communication before attempted surgical repair.

References