Management of Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery in an Adult With Ischemic Cardiomyopathy and Pulmonary Hypertension

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Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital anomaly that usually presents in infancy with cardiomyopathy and congestive heart failure. Surgical reimplantation of the anomalous left coronary artery is offered as definitive treatment in infancy. We describe the case of a 26-year-old man presenting with cardiomyopathy who was identified to have an anomalous left coronary artery from the pulmonary artery. Surgical reimplantation presented a high risk due to severe postcapillary pulmonary hypertension. Transcatheter occlusion of the proximal left main coronary artery was obtained. The patient was started subsequently on anticoagulant therapy. Surgical coronary artery bypass was performed 3-months later after resolution of postcapillary pulmonary hypertension. The patient remains well at 1-year follow-up.


A abnormal left coronary artery arising from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly that usually presents in infancy. The definitive treatment of ALCAPA is surgical reimplantation of the left main coronary artery to the aorta. Without surgical treatment, the lesion is commonly fatal within the first months of life. Occasionally, patients with ALCAPA may survive to adulthood [1–4]. Such patients present complex management challenges related to consequences of chronic myocardial ischemia [1–4]. We report the case of a 26-year-old man with ALCAPA presenting with cardiomyopathy and postcapillary pulmonary hypertension. An innovative management approach was undertaken to establish a 2 coronary artery system.

A 26-year-old man with past medical history of cardiomyopathy was referred to our clinic for evaluation. He complained of dyspnea (New York Heart Association class III) without chest pain. Physical examination revealed a regular heart rate without signs of heart failure. A 12-lead electrocardiogram was normal. Echocardiogram showed restrictive cardiomyopathy with reduced left ventricular ejection fraction (0.45) and severe pulmonary hypertension (PHT). Estimated right ventricular systolic pressure derived from the peak tricuspid regurgitant flow velocity was 80 mm Hg. Right coronary artery (RCA) appeared significantly dilated with turbulent and increased flow on color flow mapping. The origin of the left main coronary artery (LMCA) was not visualized. Multislice tomodensitometry scan demonstrated LMCA originating from the posterior aspect of the main pulmonary artery with significant dilatation of the epicardial coronary vessels. Cardiac catheterization data at presentation in room air and with inhaled nitric oxide are presented in Table 1. Severe postcapillary PHT was confirmed in the presence of severely elevated left heart filling pressure. Selective right coronary artery angiogram showed collaterals from the RCA filling the LMCA and main pulmonary artery (Fig 1). Surgical reimplantation of the LMCA in the presence of severe PHT presented a high risk. Alternatively, therapy with an afterload reducing agent for ventricular dysfunction presented the risk of myocardial ischemia. It was elected to obtain percutaneous embolization of the proximal LMCA at its origin from the PA prior to initiation of anticoagulant therapy. A 15-minute occlusion test of proximal LMCA was well tolerated; there was no evidence of myocardial ischemia on a continuous 12-lead electrocardiogram, confirming good collateralization from RCA. Percutaneous placement of a single detachable coil (MReye Embolization Coil 035-6.5-5; William Cook Europe, Bjerreby, Denmark) (Fig 2A;B) was undertaken in the proximal LMCA. Subsequently, therapy with diuretics (furosemide 40 mg/day, spironolactone 75 mg/day) and an afterload reducing agent (enalapril 20 mg/day) was initiated. Data from follow-up cardiac catheterization 3-months later is presented in Table 1. Resolution of pulmonary hypertension was noted with near normal left heart filling pressure. A myocardial scintigram for assessment of myocardial perfusion was considered at a multidisciplinary discussion; it was not pursued to avoid

Table 1. Cardiac Catheterization Data at Presentation (on Room Air and NO/O2) and After 3 Months of Anticoagulant Therapy

<table>
<thead>
<tr>
<th>Data</th>
<th>Room Air</th>
<th>NO + O2</th>
<th>s/p CHF Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aorta (mean, mm Hg)</td>
<td>82</td>
<td>89</td>
<td>60</td>
</tr>
<tr>
<td>PA (mean, mm Hg)</td>
<td>56</td>
<td>56</td>
<td>21</td>
</tr>
<tr>
<td>PCW (mean, mm Hg)</td>
<td>30</td>
<td>30</td>
<td>12</td>
</tr>
<tr>
<td>CI (L/min⁻¹/m²)</td>
<td>2.3</td>
<td>2.3</td>
<td>2.5</td>
</tr>
<tr>
<td>PVR (Wood units/m)</td>
<td>11.3</td>
<td>11.3</td>
<td>3.6</td>
</tr>
</tbody>
</table>

CHF = congestive heart failure; CI = cardiac index; NO = nitric oxide; O₂ = oxygen; PA = pulmonary artery; PCW = pulmonary capillary wedge pressure; PVR = pulmonary vascular resistance; s/p = status post.
additional radiation. We sought to obtain a 2-coronary circulation in this young patient as the cardiomyopathy was of ischemic etiology. Surgical coronary artery bypass (CAB) was performed with an internal mammary artery graft to the left anterior descending coronary artery with a complementary saphenous vein graft to the first marginal artery. Postoperative course was uneventful. At last follow-up one-year after CAB, the patient had no symptoms related to heart failure or angina. There was no evidence of PHT on Doppler echocardiography. Left ventricular function was submarginal with an ejection fraction of 0.50. The patient has failed to maintain further cardiac follow-up in spite of multiple efforts to persuade him to return for surveillance imaging to evaluate patency of the CAB graft.

Comment

ALCAPA commonly presents in infancy with dilated cardiomyopathy and congestive cardiac failure. Rarely, patients may present in adulthood with angina, dyspnea, sudden death, or ventricular arrhythmia [1–4]. Occasionally an older patient with adequate collaterals may have variable onset of symptoms [4]. Adult patients with ALCAPA are at risk for sudden death. Surgical ligation of the LMCA has been offered to such patients [5]. More recently, transcatheter embolization of the LMCA has been reported in a patient felt to be unsuitable for surgical reimplantation of the LMCA [6]. Revascularization of the myocardium by establishing a 2 coronary artery system is a preferred treatment approach for these patients [7].

Pulmonary hypertension in patients with ALCAPA is usually secondary to left ventricular diastolic dysfunction. It usually responds favorably to anticoagulopathy, improving the patient’s candidacy for surgical establishment of a 2 coronary artery system. There is, however, risk of myocardial ischemia with initiation of anticoagulopathy therapy due to coronary artery steal. In our case anticoagulopathy therapy could be administered safely after transcatheter occlusion of the LMCA, thereby eliminating coronary steal. After amelioration of postcapillary PHT, we elected to offer the internal mammary artery graft to the left anterior descending artery with a complementary
saphenous vein graft to the first marginal branch to establish a 2 coronary artery circulation [8]. It should be noted that the presence of a coil in the LMCA precluded direct reimplantation of the LMCA. Coronary artery bypass was the best treatment option in this patient.

In conclusion, we report a novel hybrid approach management that has not been previously described in a case with an unusual presentation of ALCAPA.

References