Congenital atresia of left coronary ostium

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SUMMARY A two-year-old girl, who presented with congestive heart failure and an extensive anterolateral infarction, was thought to have anomalous origin of the left coronary artery from the pulmonary artery. She improved rapidly and remained symptom free until 13 years of age, when she died suddenly. At necropsy the right coronary artery was found to be normal but in the left aortic sinus a dimple was the only remnant of the left coronary ostium. The proximal segment of the left coronary artery was diminutive and did not connect with the aortic root. This is the first report of a long term follow-up of a patient with this rare anomaly.

The majority of left coronary artery malformations are anomalies of its site of origin. The clinical picture in patients with aberrant origin of the left coronary artery from the pulmonary artery, leading to myocardial infarction and congestive heart failure in infancy, is well recognised. We describe a patient who in infancy presented with the same clinical and electrocardiographic features, became asymptomatic later on in childhood, and died suddenly at 13 years of age. At necropsy a normally situated but atretic left coronary ostium was found.

Case report

A girl was born by normal delivery after an uneventful pregnancy, birthweight 3 kg, both parents and an older sister were healthy, and growth and development were normal until 22 months of age when anorexia, tiredness, and tachypnoea were noticed. A chest x-ray film, taken because a respiratory infection was suspected, showed gross cardiomegaly and increased pulmonary vascular markings. When admitted on 24 April 1969, the patient was pale, sweating, and obviously in cardiac distress. The heart rate was 160/min and the respiratory rate was 68/min. The face was puffy and the liver was palpable 6 cm below the costal margin. On palpation a heaving left ventricular impulse, displaced to the mid-axillary line, was felt. A grade 2/6 systolic murmur and a loud gallop were heard at the apex. The electrocardiogram (Fig. 1) showed sinus rhythm and a mean frontal QRS axis of +30°. Deep Q waves and sharply inverted T waves were seen in I, aVL, and the left precordial leads. In V3 and V4 the ST segment was much raised. The diagnosis was anterolateral myocardial infarction, probably caused by anomalous origin of the left coronary artery from the pulmonary artery. After two weeks of treatment with digoxin and diuretics, the tachypnoea, tachycardia, and hepatomegaly disappeared and the patient was discharged.

She was readmitted for cardiac investigation at 2½ years of age. The apex beat was displaced and a soft systolic murmur was heard at the fourth left interscostal space. There were no signs of congestive heart failure. The electrocardiogram showed the same Q waves but the ST elevation had disappeared.

Right and left cardiac catheterisation (Table) disclosed normal pressures, with the exception of a slightly raised pulmonary capillary venous (mean 14 mmHg) and left ventricular end-diastolic pressure (12 to 18 mmHg). Oximetry showed no evidence of left-to-right shunting. Angiography of the aortic root showed absence of a left main coronary artery and a large, slightly tortuous right coronary artery. Branches of the left coronary artery were seen to fill from the right side, but the proximal segment of the left coronary artery was not seen. There was no passage of contrast material into the pulmonary artery. Though direct proof of the connection was lacking, the angiograms were thought to confirm the diagnosis of anomalous left coronary artery originating from the pulmonary artery.

The patient remained symptom free and was examined at yearly intervals. When last seen in March 1980, physical examination was unremarkable. The electrocardiogram (Fig. 1) disclosed deep but narrow Q waves and T inversion from V4 to V6. In V4 the ST segment was slightly elevated. On the chest x-ray film
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Fig. 1 The electrocardiogram at 22 months of age (A) shows deep Q waves and inverted T waves in I, aVL, V5, and V6. The ST segment is much raised in V3 and V4. At 12 years of age (B) the Q waves persist but the ST alterations are less conspicuous.

Table Haemodynamic data (at age 2 years 6 months)

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mmHg)</th>
<th>Oxygen saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>68, 67</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>a 5 v 4 m 3</td>
<td>66, 69</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>30/5</td>
<td>66, 66</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>30/15 m 23</td>
<td>67, 65</td>
</tr>
<tr>
<td>Pulmonary capillary</td>
<td>m 14</td>
<td></td>
</tr>
<tr>
<td>Left ventricle</td>
<td>120/12-16</td>
<td>99</td>
</tr>
<tr>
<td>Aorta</td>
<td>120/65</td>
<td></td>
</tr>
</tbody>
</table>

the heart was normal. The M-mode echocardiogram showed a slight increase of the end-diastolic left ventricular dimension (43 mm) and the fractional shortening of the left ventricle with systole was decreased (23%). The left atrial dimension (23 mm) and the thickness of the left ventricular posterior wall (7 mm) and interventricular septum (8 mm) were normal. Exercise testing (maximum load 600 kpm/min during 1 min) provoked no chest pain. There was no ST segment depression but the T waves in V4 to V6, which were negative at rest, became positive.

On the basis of these results it was felt that cardiac surgery and a saphenous vein graft could be postponed for a few years. The patient remained symptom free. At 13 years of age she died at home suddenly. One morning she was found pale, unconscious, and gasping. She was given mouth-to-mouth respiration and rushed to the emergency department of the hospital but was dead on arrival. The days before her death she had had a slight fever, probably unrelated to the fatal outcome.

At necropsy the right coronary artery was found to arise normally from its coronary sinus. In the left aortic sinus a dimple was the only remnant of the left coronary ostium. The proximal segment of the left coronary artery was diminutive. Before the heart was opened, a coronary angiogram with injection of the right coronary artery (Fig. 2) was performed. After

Fig. 2 Postmortem coronary arteriogram after injection of the right coronary artery. Numerous small collaterals produce retrograde filling of the left coronary artery. Its main stem, however, is short and small and does not communicate with the coronary ostium.
opacification of the large and slightly tortuous right coronary artery, the branches of the left coronary artery were seen to fill retrogradely via collaterals. Even at the point of maximum filling the branches of the left coronary artery were distinctly smaller than those of the right. The main stem of the left coronary artery was very short and narrow and did not communicate with the normally situated but atretic coronary orifice. Transverse sections of the heart showed a large \((2 \times 3 \text{ cm})\) fibrotic scar in the anterior and apical wall of the left ventricle. The entire left ventricular endocardium appeared thickened and fibrotic. The major histological finding was a zone of fresh infarction in the interventricular septum, characterised by interstitial oedema, eosinophilic coloration of the sarco plasma, and pyknosis of cell nuclei.

Discussion

Atresia of the left coronary ostium is a rare congenital anomaly. To our knowledge, five similar cases have been reported.\(^3\)\(^-\)\(^7\) The clinical picture in our case and in two previously reported infants\(^3\)\(^\,\)\(^4\) was indistinguishable from that observed in anomalous origin of the left coronary artery from the pulmonary artery. As characteristically seen in the latter condition, our patient presented with congestive heart failure, massive cardiac dilatation, and anterolateral infarction in infancy, with \(Q\) waves and \(ST\) elevation in I, aVL, and the left precordial leads.\(^1\)\(^\,\)\(^2\)

A unique feature of our case is the duration of follow-up from an episode of myocardial infarction in infancy to adolescence. Between three and 13 years of age the patient was symptom free and had a normal exercise tolerance. The cardiac volume on the chest x-ray film had become normal.

Cardiac catheterisation may contribute to the differential diagnosis if oximetry discloses a left-to-right shunt in the pulmonary artery. By contrast with older children with anomalous origin of the left coronary artery, however, infants with this anomaly rarely have a significant increase of oxygen saturation in the pulmonary artery.\(^2\) Only angiography, carefully interpreted, permits an accurate diagnosis. In our case opacification of a large right coronary artery and failure to fill the left coronary artery after aortic root injection were thought to corroborate the diagnosis of anomalous origin of the left coronary artery. The non-opacification of the main stem of the left coronary artery was attributed to poor collateral circulation.

The same mistake was made in the two reported infants.\(^3\)\(^,\)\(^5\) We therefore suggest basing the diagnosis of anomalous origin of the left coronary artery on the visualisation of the anomalous connection after retrograde filling, which is possible in most cases,\(^2\) and not only on the non-opacification of the left coronary artery after injection of the aortic root. Failure to show the left main coronary artery after retrograde filling from the right coronary artery strongly suggests atresia of the left coronary ostium.

Judging from the postmortem angiograms, the anomaly in our patient was probably not amenable to surgery. The small calibre of the main stem and major branches of the left coronary artery precluded the construction of a saphenous vein graft. Successful surgical correction and hence creation of a two coronary system have been reported in one adolescent\(^4\) and one adult\(^7\) who both presented with long standing angina pectoris.

References


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