Compression of an anomalous single coronary artery from pulmonary artery by banding

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Abstract

This report describes a case with double outlet right ventricle and doubly committed ventricular septal defect associated with congenital diaphragmatic hernia. The patient underwent arterial banding and clipping of patent ductus arteriosus after repair of the diaphragmatic hernia. At 6 months, cardiac catheterization revealed anomalous origin of a single coronary artery from the pulmonary artery of the proximal banding. Although ischaemic symptoms had not been observed, the banding had resulted in compression of the coronary ostium. An emergency Rastelli procedure with Damus–Kaye–Stansel anastomosis was successfully performed.

Keywords: Diaphragm • Congenital–cyanotic • Coronary disease

Anomalous origin of a coronary artery from the pulmonary artery is a rare congenital heart disease that can prove lethal during infancy. The most common type of this disorder is an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), occurring in 30% of all congenital coronary anomalies. Herein, we present a case with anomalous origin of a single coronary artery (SCA) from the pulmonary artery (ASCAPA), double outlet right ventricle (DORV) and doubly committed ventricular septal defect (VSD) associated with congenital diaphragmatic hernia (CDH) who underwent successful operative management.

A 2254-g male infant who was referred at 25 weeks gestation with foetal diagnosis of growth restriction, VSD and left-sided CDH was delivered by planned Caesarean section at 38 weeks gestation. Immediately after birth, the newborn was intubated, and physiological stabilization was achieved. On the second day of life, the patient underwent CDH repair. Pleural drainage and medical treatment of chylothorax were required post operatively, and the patient required mechanical ventilation for 12 days followed by nasal directional positive airway pressure support for 26 days. At 36 days, echocardiography showed DORV, doubly committed VSD, atrial septal defect (ASD), bidirectional patent ductus arteriosus (PDA) and good left ventricular function. Electrocardiogram was notable for the absence of any deep Q or ST-T changes. The patient subsequently underwent pulmonary artery banding (PAB) and clipping of PDA through median sternotomy because of high pulmonary vascular resistance. Treatment for pulmonary hypertension (PH) was administered with beraprost sodium and home oxygen therapy.

Improvement of PH was observed 6 months after PAB. Cardiac catheterization at this time revealed ASCAPA of the proximal banding. The banding tape compressed the coronary ostium, resulting in a 55% diameter reduction (Fig. 1A). The distal PA pressure and resistance were still high (Pd/Pa: 0.69; Rp: 4.2 WU). Although no ischaemic findings were present, emergency corrective surgery was performed.

Through a median re-sternotomy, the patient was placed on cardiopulmonary bypass. An infusion of cardioplegic solution was administered into the PA trunk after occlusion of the bilateral PA. The banding tape was carefully removed, and the PA trunk was transected just distal to the banding. The coronary ostium was identified without any stenosis. An SCA was visualized that arose from the left-sided posterior wall. Definitive repair without translocation of the coronary artery was proposed due to the anatomical space relation. The Rastelli procedure with Damus–Kaye–Stansel anastomosis was performed. The right ventricular outflow tract was reconstructed with a handmade bi-leaflet valved conduit (14-mm expanded polytetrafluoroethylene graft). The patient was easily weaned from cardiopulmonary bypass using nitric oxide. Echocardiography on the 22nd post operative day showed normal left ventricular function, no neo-aortic regurgitation and minor residual VSD (pressure gradient, 31 mmHg). A combination of beraprost sodium, sildenafil citrate and bosentan was administered to treat residual PH. At discharge, scintigraphy was notable for the absence of myocardial ischaemic findings.

Four months after the definitive repair, the patient underwent cardiac catheterization for assessment of the surgical results (Fig. 1B). PA pressure and resistance were 32/6/20 mmHg (Pd/Pa; 0.42) and 0.95 WU, respectively. Left ventricular function was normal. The SCA presented a normal aspect.

COMMENT

ASCAPA is an extremely rare congenital heart disease and often fatal. This anomaly is typically associated with other
cardiovascular anomalies, the most common of which are ASD, VSD, conotruncal defects (including tetralogy of Fallot and truncus arteriosus) and coarctation of the aorta [1]. The clinical course is often dependent on the number and severity of associated anomalies. ASCAPA can also occur as an isolated disease, but can still lead to early death because of severe left ventricular dysfunction or massive myocardial infarction. The presence of defects can allow perfusion of the SCA at a high oxygen saturation and pressure, and a favourable prognosis is indicated by the absence of clinical symptoms.

A literature review identified 22 cases of ASCAPA. Although the first case was reported in 1931 [2], pre-mortem diagnosis was not achieved until 1965 [3]. Almost all had ischaemic symptoms prior to diagnosis, and most were diagnosed on autopsy. Thirteen cases were diagnosed pre-mortem, and four of eight cases that underwent corrective surgery (Table 1) survived [4–6]. The present patient is a rare case of ASCAPA without ischaemic findings. Persistent PH caused by hypoplastic pulmonary vascular beds associated with CDH combined with high oxygenation in the PA through intra-cardiac defects delayed clinical symptoms and facilitated survival. In addition, the PAB had kept high pressure at the coronary ostium and not severely compressed the coronary artery.

A review of four patients who did not survive operative management revealed that the median age at the first diagnosis was 10 weeks. In two cases, pre operative thallium scintigram demonstrated a large perfusion defect. All patients died due to poor left ventricular function in the early post operative period. Autopsy revealed satisfactory coronary repair but widespread infarctions resulting in death.

In contrast, four patients who survived operative repair were diagnosed at the median age of 8 weeks. One patient after direct re-implantation from the pulmonary posterior wall required surgical repair for coronary ostium stenosis at 14 years.

Table 1: Summary of ASCAPA that underwent corrective surgery

<table>
<thead>
<tr>
<th>First author</th>
<th>Year</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Leading symptoms</th>
<th>Associated anomalies</th>
<th>Coronary distribution</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Goldblatt</td>
<td>1984</td>
<td>F</td>
<td>Angiography</td>
<td>Difficulty in feeding, tachypnoea Despnea, cough</td>
<td>Pseudo-CoA, bicuspid AV, SUA</td>
<td>Posterior wall trunk</td>
<td>Takeuchi</td>
<td>Death</td>
</tr>
<tr>
<td>Goldblatt</td>
<td>1984</td>
<td>F</td>
<td>Angiography</td>
<td>Tachypnoea, despnea</td>
<td>—</td>
<td>RT PA</td>
<td>Direct re-implantation</td>
<td>Death</td>
</tr>
<tr>
<td>Urcelay</td>
<td>1994</td>
<td>M</td>
<td>Angiography</td>
<td>Cough</td>
<td>VSD</td>
<td>Posterior sinus</td>
<td>VSD closure, direct re-implantation</td>
<td>Survival (14 years)</td>
</tr>
<tr>
<td>Urcelay</td>
<td>1994</td>
<td>F</td>
<td>Angiography</td>
<td>Heart failure, tachypnoea despnea</td>
<td>CoA</td>
<td>RT PA</td>
<td>Direct re-implantation</td>
<td>Death</td>
</tr>
<tr>
<td>Santoro</td>
<td>1995</td>
<td>M</td>
<td>Angiography</td>
<td>Heart failure</td>
<td>VSD</td>
<td>Anterior wall trunk</td>
<td>Takeuchi</td>
<td>Death</td>
</tr>
<tr>
<td>Hensch</td>
<td>1997</td>
<td>M</td>
<td>Angiography</td>
<td>Shock (p/s PAB)</td>
<td>CoA</td>
<td>Trunk</td>
<td>VSD, CoA</td>
<td>Survival</td>
</tr>
<tr>
<td>Chen</td>
<td>2006</td>
<td>F</td>
<td>Echography</td>
<td>Heart failure</td>
<td>VSD</td>
<td>Posterior wall trunk</td>
<td>Takeuchi</td>
<td>Survival</td>
</tr>
<tr>
<td>Mirza</td>
<td>2008</td>
<td>M</td>
<td>Angiography</td>
<td>—</td>
<td>VSD closure, direct re-implantation</td>
<td>Survival</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Although direct re-implantation is an ideal procedure, distortion or tenting of the coronary arteries can occur, especially in the case of an SCA origin in the posterior pulmonary wall. The alternative is the Takeuchi procedure, but post operative pulmonary stenosis, intrapulmonary baffle obstruction and leak can occur. Another strategy is the spiral-cuff technique, in which an elongated coronary artery is used for the repair of ALCAPA [8]. However, aneurysmal dilatation of the new coronary artery can also occur.

In the present case, direct re-implantation was difficult because of the SCA arising from the left-side posterior pulmonary wall. Furthermore, because of PAB, there was no surplus pulmonary wall available to use for the spiral-cuff technique. Although reoperation for the right ventricular outflow tract may be required in the future, it seemed reasonable to perform the Rastelli procedure with Damus-Kaye-Stansel anastomosis, in which coronary transfer was not necessary and in which intraventricular rerouting through the doubly committed VSD was easily performed.

Conflict of interest: none declared.

REFERENCES