Sudden Cardiac Arrest Associated with an Anomalous Aortic Origin of the Left Coronary Artery from the Opposite Sinus of Valsalva

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Abstract

A 13-year-old boy was brought to our hospital after recovering from ventricular fibrillation that occurred after an episode of chest pain during training with his soccer team. Subsequent 64-slice multidetector computed tomography revealed the left coronary artery arising from the right sinus of Valsalva, which coursed between the ascending aorta and root of the main pulmonary artery. Surgical correction including unroofing of the left coronary ostium and pulmonary artery translocation was performed successfully. One year later, he remained asymptomatic and was back on his soccer team.

Key words: sudden cardiac death, coronary artery anomaly, anomalous aortic origin of the coronary artery, surgery

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Case Report

A 13-year-old male junior high school student without any past history or familial history of cardiac disease or sudden death had been suffering from occasional chest pain during training with his soccer team. In August 2012, he felt chest pain during training, which required him to take a rest at the school infirmary, where he suddenly fell down and lost consciousness. The patient was resuscitated immediately by the school nurse with cardiac compression and an automated external defibrillator, which detected ventricular fibrillation (Fig. 1). He regained consciousness while being taken to our hospital.

Laboratory data showed no significant elevation of either the creatine kinase or troponin I levels. An electrocardiogram (ECG) showed a regular sinus rhythm without J waves, delta waves or Brugada type ST patterns, which were also not detected when using modified precordial leads. The corrected QT interval was 0.442. ECGs of the patient’s family members were also examined, which did not show any abnormal findings. Late potential and T-wave alternans were both negative. Transthoracic echocardiography revealed a normal ventricular morphology without segmental wall-motion abnormalities, or valvular disease. Blood-pool scintigraphy using technetium-99m showed normal right and left ventricular systolic functions. No late gadolinium enhancement was noted on cardiac magnetic resonance imaging, and gallium-67 scintigraphy indicated no abnormal accumulation. An epinephrine provocation test showed no significant QT prolongation. A genetic analysis of the most common gene loci (KCNQ1, KCNH2, SCN5A, KCNE1, and KCNE2) was negative for long QT syndrome.

Subsequent 64-slice multidetector computed tomography revealed an anomalous origin of the left coronary artery arising from the right coronary sinus of Valsalva (Fig. 2A). The left main coronary artery passed between the ascending artery and the root of the main pulmonary artery, which was

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Coronary angiography showed that the left coronary artery originated from the right sinus without significant stenosis, while the right coronary artery was normal. Based on these findings, we diagnosed the patient with an anomalous left coronary artery arising from the right sinus coursing between the aorta and pulmonary artery. A spasm provocation test with acetylcholine was not performed, since the catheter was incompletely engaged. However, a treadmill exercise test was performed, and the patient reached Stage IV of the standard Bruce protocol with an appropriate blood pressure and heart rate response during both exercise and recovery, ST segment and T wave changes, arrhythmias and cardiac symptoms were absent. Exercise stress technetium-99m scintigraphy was also conducted, which showed no myocardial ischemic changes.

In order to prevent sudden cardiac death, we chose surgical correction of the coronary anatomy. Following median sternotomy, unroofing of the left coronary ostium (cut-back procedure) was performed, since quite a short proximal portion of the left coronary artery remained intra-murally in the aortic wall. Pulmonary artery translocation using a vascular graft was also successfully performed, as shown in Fig. 3.

Surgical correction of the anomalous left coronary artery was completely achieved, as shown in Fig. 3, and an exercise stress ECG performed again after the operation revealed no ischemic changes; therefore, placement of an implantable cardioverter defibrillator was not planned. One year later, the patient remained asymptomatic and continued to play on his soccer team.

Discussion

The anomalous aortic origin of the coronary artery
(AAOCA) arising from the opposite sinus of Valsalva has received much attention due to its association with sudden cardiac death (SCD) in otherwise healthy individuals. Although the true prevalence of this anomaly is difficult to ascertain, previous data suggest that the rate of AAOCA is in the range of 0.1-0.3% (1-3). An anomalous right coronary artery arising from the left sinus is estimated to be 6 to 10 times more common than an anomalous left coronary artery (ALCA) arising from the right sinus (3). SCD occurs much more frequently in patients with ALCA in which the left main coronary courses between the aorta and pulmonary artery (4-6). Physical exertion is thought to significantly increase the risk of SCD in patients with AAOCAs, based on previously reported cases (Table).

The mechanism of SCD in patients with AAOCA is believed to involve episodic myocardial ischemia, according to pathological knowledge (6, 22, 23). Several theories have been developed to explain the development of myocardial ischemia in this settings, such as the effect of acute angle take-off, a resultant slit-like orifice, compression between the aorta and pulmonary artery, spasms and the intra-mural course (i.e., within the aortic wall) of the proximal portion of the anomalous coronary artery for a variable distance (6, 21, 24-26).

The majority of patients with these anomalies who died suddenly had no recognized warning symptoms. In addition, although the mechanism is uncertain, exercise stress tests do not show myocardial ischemia in the majority of affected patients (6). The results of treadmill exercise tests and exercise stress scintigraphy were normal in this case, despite the patient having sometimes suffered from chest pain during exercise. In patients with the clinical features of AAOCA, the use of surgical coronary revascularization in all cases of ALCA coursing between the aorta and pulmonary artery regardless of evidence of ischemia is supported by a consensus statement in the Guidelines for the Management of Adults with Congenital Heart Disease (27).

There are multiple surgical options due to the wide spectrum of variation in coronary morphology (28). Bypass grafting has become less frequently used in recent years as a result of early graft failure (29). Other approaches include pulmonary artery translocation (30), reimplantation of the anomalous vessel into its appropriate sinus (4, 31), patch augmentation (32), and unroofing (33). Although some case reports on the use of stents to manage AAOCAs in adults have been published (7, 17), data regarding the long-term prognosis remain scant, and, moreover, stents have limited application in growing children.

This report describes a successful case of recovery from sudden cardiac arrest associated with AAOCA that was repaired with surgical vascularization. Awareness of this rare

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**Table.** Previously Reported Cases of Anomalous Aortic Origin of the Coronary Artery from Opposite Sinus from 2000 to 2012

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age</th>
<th>Gender</th>
<th>Anomaly</th>
<th>Symptoms</th>
<th>Triggered situation</th>
<th>Treatment</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>51 M</td>
<td>M</td>
<td>ARCA</td>
<td>CP</td>
<td>Exertion</td>
<td>Stent</td>
<td>Alive</td>
</tr>
<tr>
<td>7</td>
<td>45 M</td>
<td>M</td>
<td>ARCA</td>
<td>CP</td>
<td>Exertion</td>
<td>Stent</td>
<td>Alive</td>
</tr>
<tr>
<td>8</td>
<td>10 F</td>
<td>F</td>
<td>ALCA</td>
<td>SCA</td>
<td>None (resting)</td>
<td></td>
<td>Sudden death</td>
</tr>
<tr>
<td>9</td>
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<td>M</td>
<td>ALCA</td>
<td>Dyspnea</td>
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<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
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<td>M</td>
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<td>SCA</td>
<td>Running</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
<td>11</td>
<td>18 M</td>
<td>M</td>
<td>ALCA</td>
<td>SCA</td>
<td>Running</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
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<td>56 M</td>
<td>M</td>
<td>ARCA</td>
<td>CP</td>
<td>Exertion</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
<td>12</td>
<td>33 M</td>
<td>M</td>
<td>ARCA</td>
<td>CP</td>
<td>Exertion</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
<td>13</td>
<td>18 M</td>
<td>M</td>
<td>ALCA</td>
<td>CP, syncope</td>
<td>Exertion</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
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<td>15 M</td>
<td>M</td>
<td>ALCA</td>
<td>CP, syncope</td>
<td>Playing soccer</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
<td>15</td>
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<td>M</td>
<td>ALCA</td>
<td>CP</td>
<td>Exertion</td>
<td>Surgery</td>
<td>Alive</td>
</tr>
<tr>
<td>16</td>
<td>24 M</td>
<td>M</td>
<td>ARCA</td>
<td>SCA</td>
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</tr>
<tr>
<td>17</td>
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<td>ARCA</td>
<td>CP</td>
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<td>Stent</td>
<td>Alive</td>
</tr>
<tr>
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<td>M</td>
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<td>SCA</td>
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<td>SCA</td>
<td>General anesthesia</td>
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<td>Alive</td>
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</table>

anomaly as an important cause of SCD is important, and better knowledge is required to determine the surgical indications.

The authors state that they have no Conflict of Interest (COI).

References