Congenital pericardial defect with ruptured acute type A aortic dissection

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Abstract

We report 2 cases of congenital pericardial defect with ruptured acute type A aortic dissection. Case 1: An 83-year old man presented with sudden chest and back pain, and computed tomography (CT) showed acute aortic dissection with left pleural massive effusion. Because of his unstable haemodynamic condition with low blood pressure, an emergency operation was performed. We observed small amounts of bloody pericardial effusion, massive left-sided bloody pleural effusion and a partial left-sided pericardial defect of the pulmonary artery. The ascending aorta was replaced. The postoperative course was uneventful. Case 2: A 79-year old man presented with fainting followed by cardiac arrest and was resuscitated. Chest CT showed acute aortic dissection and massive haemothorax. Emergency operation was attempted, but was given up. We observed partial left-sided pericardial defect of the pulmonary artery. Further, we reviewed 6 cases of congenital pericardial defect with ruptured acute aortic dissection, including our 2 cases. In all the cases, the patients did not develop cardiac tamponade but had massive haemothorax due to congenital pericardial defects leading to confusion in diagnoses and surgical strategies. Therefore, it may be necessary to consider congenital pericardial defects before performing an operation in case of acute type A aortic dissection with massive haemothorax.

Keywords: Congenital pericardial defect • Ruptured acute aortic dissection • Cardiac tamponade • Left haemothorax

INTRODUCTION

Congenital pericardial defect is a rare anomaly. Previous papers report that the frequency is \( \sim 1 \) in every 10 000 autopsies and roughly 1 of 14 000 births [1]. Most cases are asymptomatic and some cases are found incidentally during cardiac surgery. If ruptured type A acute aortic dissection is presented in a patient with undiagnosed congenital pericardial defect, understanding the precise diagnosis would be impossible, leading to possible failure of treatment. We report very rare cases of ruptured type A acute aortic dissection with congenital pericardial defects.

CASE 1

An 83-year old man presented at a nearby hospital with sudden chest and back pain. The patient was diagnosed with acute Stanford type A aortic dissection by enhanced computed tomography (CT) and was referred to our hospital immediately.

On examination, the patient was conscious, with blood pressure of 96/71 mmHg and the pulse was 89 beats/min.

Chest radiography showed an enlarged mediastinal shadow and lowered density in the left lung field. Enhanced chest CT showed aortic dissection confined to the ascending aorta and a massive left pleural effusion (Fig. 1a). Pericardial effusion was almost absent.

Because his haemodynamic status was unstable with low blood pressure, the patient was taken to the operating room for acute Stanford type A aortic dissection associated with unexplained left thoracic bleeding.

After median sternotomy, extracorporeal circulation was established with perfusion via the left common femoral artery, and right atrium drainage was performed. When the pericardium was opened, a small amount of bloody pericardial effusion was aspirated. Following this, we noticed that there was a 30 × 30 mm pericardial defect on the left side of the pulmonary artery. We drew 1000 ml of blood from the left pleural cavity (Fig. 1c). The ascending aorta was replaced with a prosthetic graft under deep hypothermic circulatory arrest with antegrade selective cerebral perfusion. The pericardial defect was not repaired.

The postoperative course was uneventful, and he was discharged on the 12th postoperative day in stable condition.

CASE 2

A 79-year old man was transported to a local hospital because of syncope. On arrival, he went into cardiac arrest and was resuscitated. Plain chest CT showed an enlarged ascending aorta with a calcified intimal flap indicating acute aortic dissection and massive haemothorax (Fig. 1c). He was referred to the emergency department of our hospital. On arrival, he had another cardiac arrest and
was immediately transferred to the operating room while undergoing heart massage. After median sternotomy, massive bleeding around the ascending aorta was noticed. We tried to resuscitate him but were unable to regain heart beat. We observed that there was a 30 × 40 mm pericardial defect on the left side of the pulmonary artery and massive left haemothorax. The blood from the dissection had spilled into the left thoracic cavity via the pericardial defect, which caused critical hypovolaemic shock.

**DISCUSSION**

Congenital pericardial defect is an uncommon anomaly, and is most often asymptomatic, but the patients sometimes complain of chest pain, dyspnoea, palpitation or dizziness [1, 2]. Congenital pericardial defect may be either partial or complete, and is usually located on the left side (86%). Patients with partial pericardial defects may encounter cardiac herniation or torsion, causing sudden shock or death. Most congenital pericardial defects are incidentally discovered during cardiac surgery [2]. A diagnosis of congenital pericardial defect is suspected if the cardiac chamber is displaced leftward on chest radiography or CT; however, in most cases, it is quite difficult to confirm the diagnosis preoperatively.

In type A acute dissection, most cases present with haemodynamic instability caused by cardiac tamponade. These findings are easily diagnosed by CT or echocardiography. However, acute type A dissection associated with haemothorax and with little pericardial effusion indicates the existence of a pericardial defect or rupture of the descending aorta. If type A acute dissection associated with rupture of the descending aorta is suspected, extended ascending to descending aorta replacement, which is quite an invasive procedure, should be performed.

There are only 6 reported cases, including our 2 cases, of congenital pericardial defect associated with ruptured acute type A dissection. Table 1 shows the characteristics of these patients.

**Table 1: Characteristics of 6 patients with ruptured acute aortic dissection associated with congenital pericardial defect**

<table>
<thead>
<tr>
<th>No.</th>
<th>Author</th>
<th>Journal, year</th>
<th>age (yr), sex</th>
<th>CPD (size, site)</th>
<th>Type of CPD</th>
<th>Preoperative status</th>
<th>Operative procedures</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Nakajima et al.</td>
<td>Ann Thorac Surg, 2004</td>
<td>84, Male</td>
<td>30 × 50 mm, left side of PA</td>
<td>Partial</td>
<td>Little pericardial effusion, massive left haemothorax</td>
<td>Ascending aorta replacement</td>
<td>Prolonged hospital stay due to CI</td>
</tr>
<tr>
<td>B</td>
<td>Matsuda et al.</td>
<td>Ann Thorac Surg, 2004</td>
<td>64, Male</td>
<td>25 × 30 mm, left side of PA</td>
<td>Partial</td>
<td>Little pericardial effusion, massive left haemothorax</td>
<td>Total arch replacement</td>
<td>Discharge on 46th POD</td>
</tr>
<tr>
<td>C</td>
<td>Nisanoglu et al.</td>
<td>Tex Heart Inst J, 2005</td>
<td>46, Male</td>
<td>Left side of heart</td>
<td>Complete</td>
<td>Little pericardial effusion, massive left haemothorax, shift of heart into the left side of thoracic cavity</td>
<td>Hemiarch replacement</td>
<td>Discharge on 7th POD</td>
</tr>
<tr>
<td>D</td>
<td>Huang et al.</td>
<td>Chirurgia, 2007</td>
<td>67, Male</td>
<td>40 × 60 mm, lower portion of pericardium</td>
<td>Partial</td>
<td>Little pericardial effusion, massive left haemothorax</td>
<td>Ascending aorta replacement and aortic valve repair</td>
<td>Discharge on 20th POD</td>
</tr>
<tr>
<td>Case 1</td>
<td>Our case</td>
<td></td>
<td>83, Male</td>
<td>30 × 30 mm, left side of PA</td>
<td>Partial</td>
<td>Little pericardial effusion, massive left haemothorax</td>
<td>Ascending aorta replacement</td>
<td>Discharge on 12th POD</td>
</tr>
<tr>
<td>Case 2</td>
<td>Our case</td>
<td></td>
<td>79, Male</td>
<td>30 × 40 mm, left side of PA</td>
<td>Partial</td>
<td>Little pericardial effusion, massive left haemothorax</td>
<td>Attempt</td>
<td>Operative death</td>
</tr>
</tbody>
</table>

CPD: congenital pericardial defect; PA: pulmonary artery; BP: blood pressure; PR: pulse rate; POD: postoperative day; CI: cerebral infarction.
dissection [1, 3–5]. The characteristics of these 6 cases are listed in Table 1. The clinical feature of congenital pericardial defect associated with ruptured acute type A dissection is similar in all 6 cases. Symptoms such as syncope were related to hypovolaemic shock due to bleeding into the thoracic cavity. Clinical imaging findings showed acute aortic dissection and massive left haemothorax with little pericardial effusion. A preoperative diagnosis of pericardial defect was suspected in only one case with a complete pericardial defect.

It is necessary to consider the possibility of a congenital pericardial defect before performing an operation in the case of acute type A aortic dissection with a massive haemothorax and little pericardial effusion.

Conflict of interest: none declared.

REFERENCES