Case Reports

Complete Left-Sided Absence of the Pericardium

in Association with Ruptured Type A Aortic Dissection Complicated by Severe Left Hemothorax

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We report an unusual clinical presentation of an acute type A aortic dissection as a left hemothorax in a patient with a congenital pericardial defect. Although the pericardial defect was diagnosed preoperatively, we could not exclude the possibility of a ruptured descending aorta until we discovered the site of the rupture during operation.

The presence of a pericardial defect would at first appear to be a fatal disadvantage in such a situation as this, due to massive bleeding into the pleural space; but we believe that in our patient spontaneous drainage of blood into the pleural cavity prevented severe cardiac tamponade. The only reason for his deteriorating hemodynamic status was hypovolemia, which was corrected with volume replacement. ***(Tex Heart Inst J 2005;32: 241-3)***

Complete left-sided absence of the pericardium is a very rare cardiac malformation,[1] and acute type A aortic dissection complicated by rupture requires prompt surgical intervention in order to prevent death from intrapericardial hemorrhage.[2] Herein, we offer an unusual clinical presentation of acute type A aortic dissection manifesting as a massive left hemothorax, which was caused by rupture of the ascending aorta into the pericardial cavity and drainage into the left hemithorax through the left-sided pericardial defect.

Case Report

A 46-year-old man diagnosed with an acute type A aortic dissection was referred to our emergency department for surgical treatment in May 2004. He had been diagnosed at another hospital by magnetic resonance imaging. He presented with acute onset of chest pain and syncope. He had a long history of atypical chest pain and hypertension. His blood pressure was 120/70 mmHg, with a heart rate of 100 beats/min. Initial evaluation of the patient included a chest radiograph, which showed mild enlargement of the mediastinal shadow and levoposition of the heart (Fig. 1A).

During surgical preparation in the intensive care unit, the patient's clinical situation suddenly worsened, and he went into shock. After substantial volume replacement, his hemodynamic condition recovered. We immediately performed bedside echocardiography to determine whether acute aortic insufficiency or cardiac tamponade due to rupture had led to this episode.

Echocardiography showed neither aortic valve disease nor pericardial tamponade. Myocardial functions were also normal, but a small amount of pericardial and a large amount of left pleural effusion were detected. On the basis of the patient's hemodynamic stability after treatment with volume replacement, contrast computed tomography (CT) was performed. The CT scan revealed a Stanford type A aortic dissection, shift of the heart into the left hemithorax, and a severe left-sided hemothorax. Our initial diagnosis was rupture of the distal aortic arch or the descending aorta into the left pleural space. However, the levoposition of the heart created suspicion that a pericardial defect had led to the left hemothorax. Careful reevaluation of the CT scan revealed a pericardial defect (Fig. 1B). Although there was still a possibility of descending aortic rupture, the patient underwent emergent surgical intervention.

Key words: *Aneurysm, dissecting; aortic aneurysm, thoracic; aortic rupture/surgery; cardiac tamponade; diagnosis, differential; hemothorax/etiology; pericardial effusion; pericardium/abnormalities*

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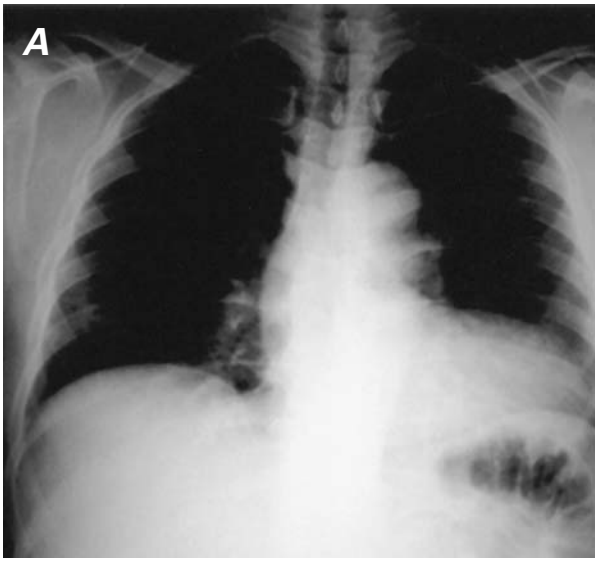
After cannulation of the right axillary artery, we performed a median sternotomy. We then noticed that the heart and aorta were not completely covered with pericardium (Fig. 2). There was a small hematoma adjacent to the ascending aorta. We established cardiopulmonary bypass by right atrial cannulation. The heart was in the left chest, where we encountered a complete absence of left-sided pericardium. We found the rupture site at the ascending aorta and evacuated 2,000 mL of blood from the left hemithorax. Intraoperative exploration revealed an intimal tear at the aortic arch that extended retrogradely toward the proximal ascending aorta. The ascending aorta and hemiarch were replaced with a supracoronary prosthetic graft. Antegrade selective cerebral perfusion was used to provide cerebral protection. The pericardial defect was left without closure. The postoperative course was uneventful, and the patient was discharged on the 7th postoperative day in stable condition.



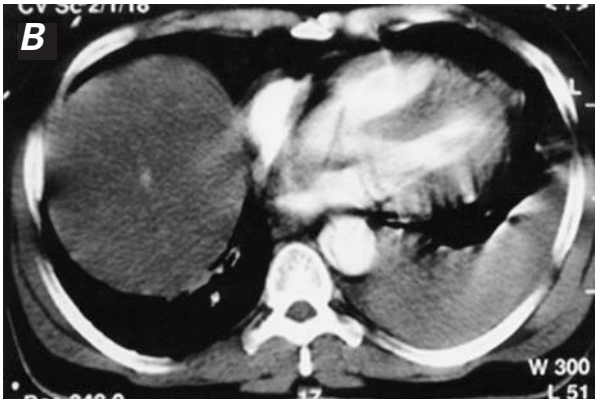
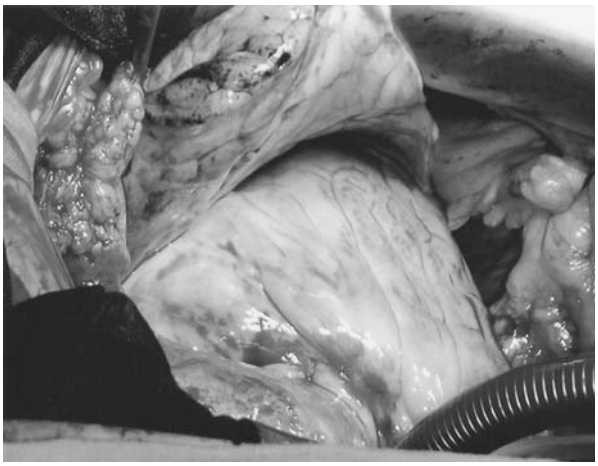


Fig. 1 ***A****) Chest radiograph shows mild enlargement of the mediastinal shadow and levoposition of the heart.* ***B****) Computed tomogram shows dissection of the descending aorta, shifting of the heart into the left hemithorax, and a severe left-sided hemothorax.*



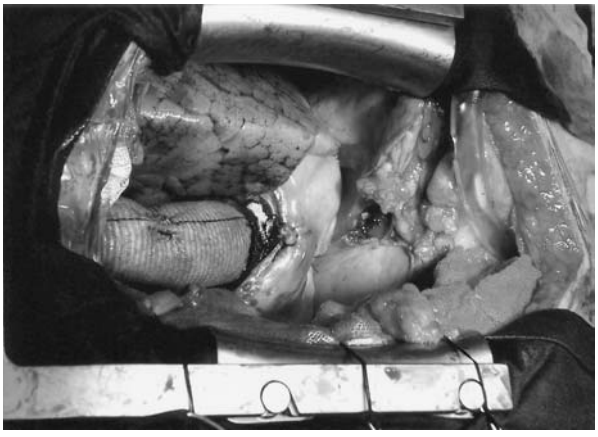


Fig. 2 *Operative photos demonstrate the complete absence of the left pericardium.*

Discussion

Type A aortic dissection is a true cardiovascular emergency that requires prompt diagnosis and surgical intervention. The cause of hemothorax in cases of ruptured type A aortic dissection is not easily determined, because of difficulty in identifying the site of the rupture. If the dissection involves the descending aorta and if the rupture site lies in that part of the aorta, the surgical strategy—including the surgical approach and establishment of extracorporeal circulation—is more complex than that for rupture of the ascending aorta.

Diagnosis of a congenital pericardial defect (CPD) may be very difficult to establish, particularly in an emergency situation such as that brought about by acute type A aortic dissection. Such a defect is usually discovered inadvertently, during operations or postmortem.[1] Definitive diagnosis can be made by CT or magnetic resonance imaging (MRI).[1,3] Various types

of CPDs exist, ranging from a small foramen to complete one-sided or totally absent pericardium. Foraminal defects have the potential to cause herniation of the left atrium or both ventricles and constriction of the coronary arteries by the pericardial rim.[1,3,4] If the pericardium is totally absent or a complete left-sided pericardial defect exists, the condition may pose a risk for dissection as a consequence of increased heart mobility and lack of pericardial attachments, particularly during chest trauma such as high-impact deceleration injury.[5] In our patient, there was no history of trauma, and dissection was probably secondary to hypertension.

In reviewing the English-language literature, we were able to find only a few cases of CPD in association with a ruptured acute type A dissection. Even though our case displays some similarities with published cases,[6,7] the case is different with respect to the type of pericardial defect and the extent of aortic dissection. The existing literature describes foraminal pericardial defects in association with ruptured type A aortic dissection that was confined to the proximal aorta—the descending aorta was not involved. Our case presents a complete left-sided pericardial defect with type A dissection involving the descending aorta. It is conceivable that the left hemothorax may have been caused by rupture of the descending aorta. The surgical strategy in treating the descending aorta—including the surgical approach and establishment of cardiopulmonary bypass—is different from that in treating the ascending aorta. Although the cause of the left hemothorax in this patient was not clearly identified preoperatively, we performed an emergency operation via a median sternotomy, on the basis of recent reports.[5-7] Intraoperatively, we noticed a complete left-sided pericardial defect and discovered the rupture of the ascending aortic root. These observations answered the question of how a ruptured acute type A aortic dissection could manifest itself in the form of a massive left hemothorax. The existence of the ascending aortic rupture certainly facilitated our surgical treatment, but the possibility that a descending aortic rupture had caused severe hemothorax was still present when the patient underwent operation. In this complicated situation, an additional thoracotomy, combined with sternotomy, may be an alternative approach to reach the ruptured descending aorta.

In the confines of the pericardial cavity, the accumulation of blood and other fluid in the pericardial space may lead to cardiac tamponade and occlusion of coronary arteries; in addition, there may be hypovolemia due to bleeding.[2] Tan and colleagues[8] reported that draining of the pericardial cavity in acute type A dissection with tamponade appeared to be a protective factor associated with decreased hospital mortality. On the other hand, in several other reports,[7,9] authors did not recommend pericardiocentesis. In our patient, the presence of a pericardial defect would at first appear to be a fatal disadvantage, due to massive bleeding into the pleural space; but we believe that spontaneous drainage of blood into the pleural cavity prevented severe cardiac tamponade, which worsens the hemodynamic status of the patient. The only reason for our patient's deteriorating hemodynamic status was hypovolemia, which resulted from direct extravasation and was corrected with volume replacement; cardiac functions were unaffected.

As shown in our case, an acute type A aortic dissection can coexist with CPD, and the patient can present with a massive left hemothorax without descending aortic rupture. The pericardial defect can make the clinical situation more complex in patients with hemorrhage due to rupture of the great vessels or cardiac chambers. The conflicting clinical presentation can delay diagnosis and surgical intervention, which of course can be fatal. Careful observation of the CT or MRI scan can in some instances reveal the pericardial defect before operation. A high index of suspicion and better understanding of this condition preoperatively will facilitate surgical management.

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