We preferred to initiate femorofemoral bypass first, to avoid risk of rupture of the giant aneurysm during sternotomy, because it was seen abutting the sternum on the lateral chest roentgenogram. The opening of the afferent artery into the aneurysm and the fistulous opening into the right atrium were closed from within the aneurysmal sac and the right atrium, respectively, to avoid risk of recurrence. On histopathologic examination, the excised wall, surprisingly, revealed cystic medial change.

We conclude that in patients with giant CAA with fistula, surgical repair is the treatment of choice and the surgical strategy should be carefully planned and individualized.

References


Annuloaortic Ectasia in a Patient With Congenital Absence of the Left Pericardium

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We report a patient with congenital absence of the left pericardium with development of progressive annuloaortic ectasia and aortic insufficiency during a 12-year period. The patient was treated with a Bentall procedure. Pathologic examination of the aorta revealed cystic medial necrosis. The surgical management and a possible association between congenital absence of pericardium and Marfan syndrome are discussed.


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A 48-year-old man diagnosed with congenital absence of the pericardium (CAP) 12 years earlier [4] presented with dyspnea upon exertion and increased neck pulsations. A type A aortic dissection had occurred in the patient’s brother when he was in his early 40s.

The physical examination revealed an increased pulse pressure and a diastolic murmur. Echocardiography demonstrated severe aortic insufficiency and annuloaortic ectasia. The sonographic windows were poor. Cardiac magnetic resonance imaging confirmed severe aortic insufficiency, with a regurgitant fraction of 50%. The aortic valve was trileaflet. The left ventricular ejection fraction was 0.45, and the left ventricle dimensions were 76 mm in end-diastole and 47 mm in end-systole. As expected, there was displacement of the heart into the left chest, with the apex pointing posteriorly.

Computed tomography angiography showed that the aortic annulus, the sinuses of Valsalva, and the sinotubular junction were aneurysmal (Fig 1). The arch and descending aorta were normal in diameter.

We recommended surgical intervention based on the presence of severe symptomatic aortic regurgitation with decreased left ventricular ejection fraction and dilated left ventricle and the presence of annuloaortic ectasia. Upon sternotomy, the heart was covered by the right lung. No adhesions were present. The left phrenic nerve was located anteriorly to the right atrioventricular groove. The aorta, pulmonary artery, and superior and inferior vena cava were in normal position. The location of the aortic sinuses and coronary ostium was normal. The ventricular cavities were displaced toward the left and posteriorly. There was a free pericardial edge next to the right atrium covering the right phrenic nerve.

Cardiopulmonary bypass was instituted by cannulating the distal ascending aorta and the right atrium. The ascending aorta, sinuses of Valsalva, and aortic leaflets were excised. Buttons containing the coronary arteries were fashioned. A Bentall procedure was performed with a 32-mm valved conduit. Histologic examination of the ascending aortic wall demonstrated medial necrosis.
Fig 1. (A–E) Computed tomography angiography demonstrates extreme displacement of the heart into the left hemithorax with the apex pointing posteriorly. The findings are consistent with congenital absence of pericardium. Note the relatively normal location of the aortic root, ascending aorta, and arch. (F) The aortic root was dilated. The aortic annulus was 3.3 cm. The sinuses of Valsalva measured 4.3 cm × 4.2 cm × 4.0 cm (z score = 2.07). The sinotubular junction was 3.8 cm. The ascending aorta was 3.9 cm × 3.6 cm. The proximal arch was 3.4 cm, and the descending aorta was 2.5 cm. (G) Histologic examination of the aortic wall was consistent with medial necrosis. Excess glycosaminoglycan deposition within the media of the aorta is seen as light blue staining on hematoxylin and eosin stained sections (magnification, ×100). (AoR = aortic root; Dao = descending aorta; LV = left ventricle; RV = right ventricle.)

The patient recovered well and is doing well 1 year after the procedure.

Comment

CAP is uncommon. It is usually diagnosed as an incidental finding in patients being evaluated for chest pain or other cardiac symptoms [1]. Coronary artery bypass grafting and ascending aorta replacement for type A dissection have been reported in these patients [2, 3, 5, 6]. Cardiac operations in these patients can be challenging because of the abnormal location of the cardiac structures. Particular attention should be paid to the location of the left phrenic nerve, which is usually located anterior to the heart and the great vessels.

During coronary artery bypass grafting, measures should be taken to prevent kinking of the grafts, particularly the left internal mammary artery. The venous grafts to the lateral wall coronary vessels need to be longer because the heart is displaced posteriorly and laterally.

Aortic root replacement and the anatomy of the aortic root in the setting of CAP with extreme left and posterior rotation of the heart have not been well described. The aortic root in this patient had the usual anatomic configuration, and the locations of the coronary and non-coronary sinuses, coronary ostia, and coronary artery origins were normal. Arterial and venous cannulation, retrograde cardioplegia cannula, left ventricular vent, and root vent placement were conducted routinely. The left phrenic nerve needed to be mobilized laterally because it was running in front of the aorta, right atrial appendage, and right ventricle.

CAP occurs as an isolated defect in most patients. However, it can be associated with cardiac or extracardiac
defects in 30% of patients [1]. A link between CAP and aortic connective tissue disorders is not established, but two previous studies have reported that association [7, 8]. In addition, several cases of aortic dissections have been described in these patients. The current patient had a family history of aortic dissection at a young age, and progressive dilatation of the aortic root had occurred during a 12-year period. Histologic findings were similar to those in patients with connective tissue disorder.

Medial necrosis is the histologic hallmark of patients with Marfan syndrome. It is also commonly seen in other groups who present with aortic dissections or aneurysms [6]. Non-Marfan patients with medial necrosis are at high risk of late adverse aortic events and should be aggressively monitored and treated to avoid acute aortic syndromes in other territories [8]. This patient did not meet the Gent criteria for the diagnosis of Marfan syndrome [9], but his family history, presentation, and histologic assessment were highly suggestive of Marfan syndrome or another aortic connective tissue disorder. Lifetime surveillance is recommended for aortic dilatation.

In conclusion, the aortic root anatomy in patients with CAP is preserved. Therefore, aortic root replacement can be safely performed without the need for technical modifications. Surgeons should be aware of the abnormal anterior location of the left phrenic nerve. CAP may be associated with abnormalities of the aortic wall that predispose patients to aortic aneurysm or dissection. Patients with CAP should be screened for root and ascending aorta aneurysms. Additional studies are necessary to further define this association.

References

Left Ventricular Assist Device and Heart Transplantation in Hemophilia A Patient
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We report here a hemophilia patient who was bridged with a left ventricle assist device and later received heart transplantation. Preparation for surgery with factor VIII supplementation, intraoperative conduct of surgery, and challenges of postoperative course are described with a brief literature review.


Hemophilia is a rare X-linked autosomal recessive disorder. Type A, the more common form, is characterized by decrease in levels of clotting factor VIII. Severity of bleeding disorder is based on circulating factor VIII levels. Levels of greater than 5%, 1% to 5%, and less than 1% are termed mild, moderate, and severe disease, respectively [1]. Patients with mild or moderate disease often go undiagnosed or are diagnosed after significant unexpected bleeding from surgery [2]. Of the estimated 400,000 patients with hemophilia worldwide, only a third will get diagnosis in their lifetime. Recombinant factor VIII has significantly improved survival of patients with hemophilia. Diabetes and hypertension, the major risk factors for cardiac disease, are twice as common in patients with hemophilia [3]. Due to improved survival, more patients with hemophilia are expected to present with cardiovascular illnesses including advanced heart failure.

We report a case of a patient hemophilia A, who presented with advanced heart failure requiring placement of left ventricular assist device (LVAD; HeartMate II, Thoratec Corp, Pleasanton, CA), aortic valve replacement and coronary artery bypass surgery as a “bridge” to heart transplantation. Institution Review Board gave authorization to report this case.

A 63-year-old man with advanced ischemic cardiomyopathy was evaluated for heart transplantation. Echocardiogram showed severe biventricular failure, moderate aortic valve regurgitation, and severe mitral and moderate tricuspid valve regurgitation. Heart catheterization documented multivessel coronary artery disease, elevated pulmonary artery pressures, and