

Off-Pump Coronary Artery Bypass in a Patient with Congenital Absence of the Left Pericardium and an Extremely Left-Rotated Heart

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ABSTRACT

Congenital anomalies of the pericardium are extremely rare and often difficult to diagnose. We describe a 66-year-old woman presenting with multivessel coronary artery disease and right heart insufficiency complicated by an extremely left-rotated heart due to a congenital absence of left pericardium. A multidetector row computerized tomographic study was performed to evaluate the torsion of the heart as well as the physical relationship between the left internal thoracic artery and the left coronary system. Off-pump coronary surgery was scheduled. Both the operative and postoperative courses were uneventful and the patient was discharged after 11 days.

BACKGROUND

Congenital pericardial defects are rare and diagnosis in adulthood is unusual because most patients with absence of the pericardium are asymptomatic [Van Son 1993]. Predominantly pericardial defects appear partially on the left thoracic pleuropericardium [Gatzoulis 2000]. Diagnosis is usually incidental or related to cardiac surgery performed for other causes. Although pericardial defects are rarely of clinical relevance, significant concern may arise from potential life-threatening complications, such as left ventricle torsion with herniation or myocardial necrosis [Van Son 1993; Brulotte 2007]. Pericardial defects that are symptomatic are commonly associated with chronic chest pain, which may make the diagnosis and therapy difficult in patients presenting with concomitant coronary artery disease. We report the case of a 66-year-old patient admitted to our department with congenital absence of left thoracic pericardium resulting in a total left displacement of the heart, right heart insufficiency, and concomitant severe coronary artery disease.

CASE REPORT

A 66-year-old female patient with a history of right heart insufficiency with moderate tricuspid valve regurgitation,

chest discomfort, and recurrent angina symptomatic for 1 month was referred to our department for coronary by-pass surgery.

The patient's chest discomfort had started 6 weeks after a surgical procedure to extirpate an abdominal cyst. The patient presented postoperatively with chest discomfort and dyspnoea. The vital signs included a blood pressure of 150/80 mm Hg and a heart rate of 75 beats/min. Physical examination revealed no remarkable findings. Electrocardiography showed a sinus rhythm with 70 beats/min, as well as R-attenuation from V1 to V5. A chest x-ray film showed cardiomegaly with a protruding left ventricular wall, displacement of the heart to the left lateral wall, and a prominent right atrium. Transthoracic echocardiography showed an unspecific left displacement of the heart and dilatation of both atria as well as dilatation and hypertrophy of the right ventricle. Moderate tricuspid valve regurgitation was also observed. The left ventricular function was lightly reduced at this time. A multidetector row computerized tomography (CT) was performed to rule out pulmonary embolism. The CT showed no signs of pulmonary embolism. However, an intrathoracic complete displacement of the heart with left rotation was revealed (Figure 1A). A left-posterior rotation of the left anterior descending coronary artery (LAD) was also revealed (Figure 1B). To further assess the ventricular function and the valve status a transesophageal echocardiography was performed. It confirmed the previous findings and also showed a left-to-right atrial shunt without hemodynamic consequences. Specific coronary diagnostic procedures were performed. Cardiac catheterization from the standard right femoral Judkins approach showed a normal-sized left ventricle with reduced global and segmental contractility, although the left-ventricular end-diastolic pressure remained within the normal range. Right and left heart catheter revealed a reduced cardiac index of 2.2 L/min per m² as well as a dilated right atrium and ventricle with altered function and mild tricuspid insufficiency. Pulmonary pressure at systole was 30 mm Hg and at diastole 4 mm Hg, middle 13 mm Hg. In addition, the atrial-septal defect with a minimal left-right shunt was confirmed.

Catheterization also demonstrated 3-vessel coronary disease. There was evidence of several proximal and medial lesions of the LAD, circumflex, and right coronary arteries. Angioplasty and stent deployment were performed unsuccessfully on the LAD lesion. The patient was scheduled for coronary artery bypass grafting. We restudied the CT

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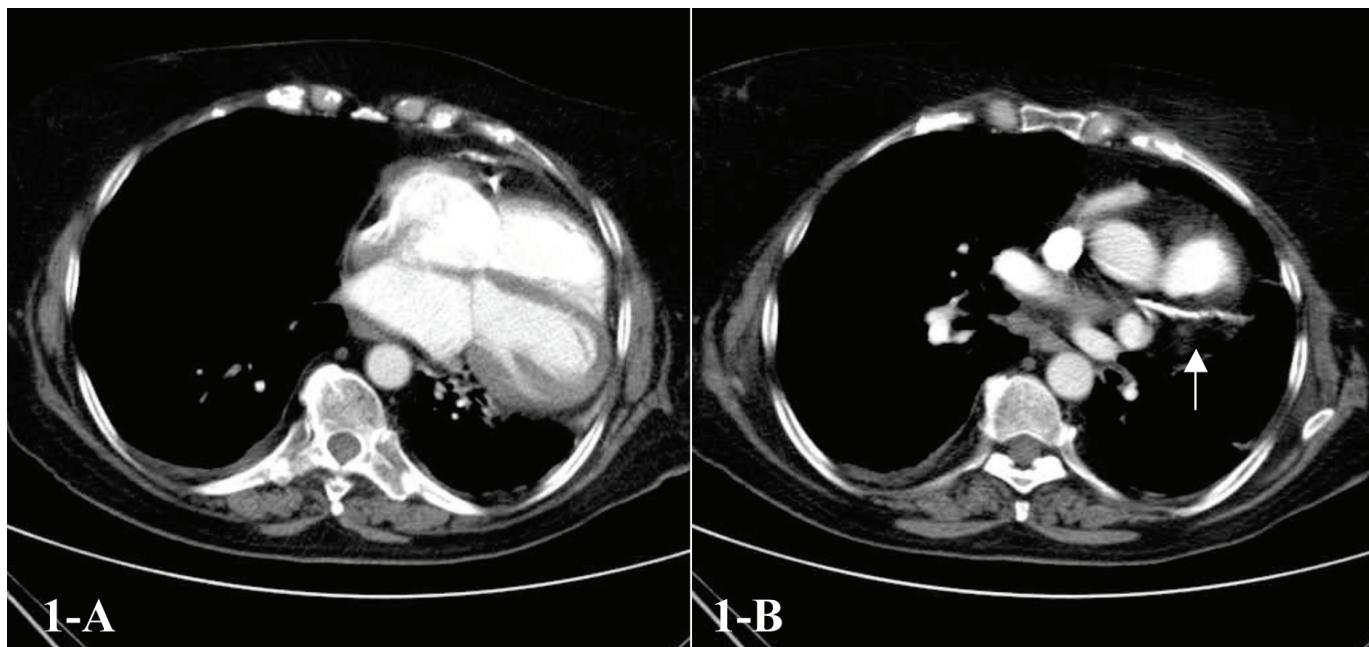


Figure 1. A, Computed tomography of the heart showing the extreme intrathoracic displacement with left posterior rotation of the heart. B, The posterior lateral position of the left anterior descending artery can be observed (arrow).

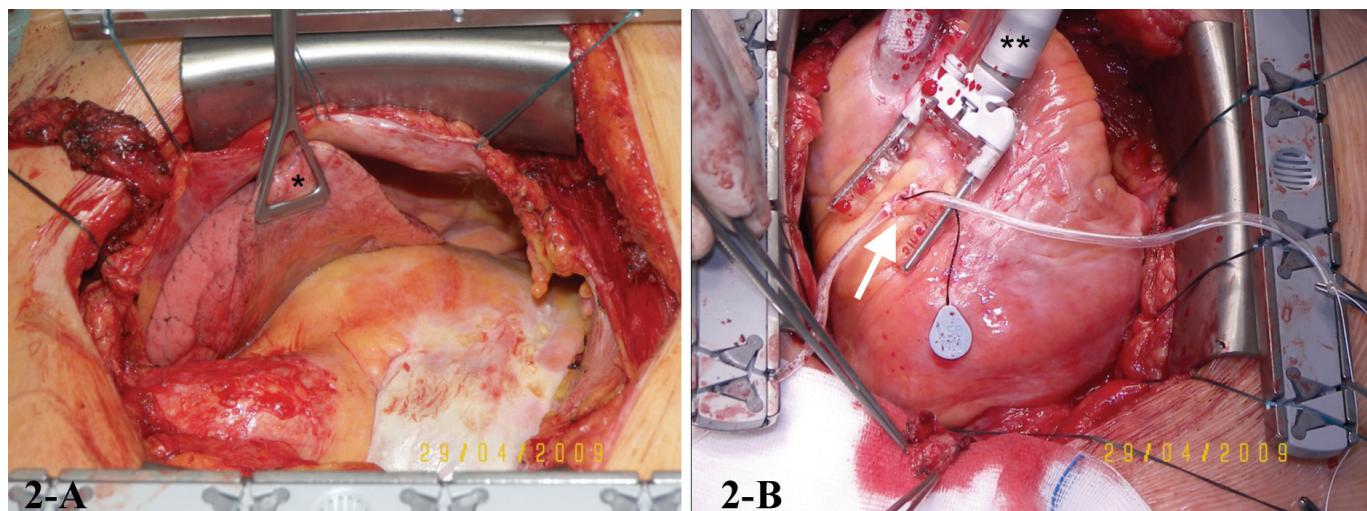


Figure 2. A, Surgeon's perspective shows the right upper lobe of the lung prolapsing into the mediastinum and held by the Duval lung forceps (*). No pericardium could be found on the left side. B, Anesthetist's perspective shows the heart rotated by a heart positioner and the tissue stabilized. The left anterior descending artery is snared by 2 vessel loops and a shunt (1.5 mm) is inserted (arrow).

findings to assess the length of the left internal thoracic artery (LITA) as an arterial graft for the LAD, and the results were satisfactory.

We performed off-pump coronary artery bypass grafting by using the LITA to the LAD artery and a sequential saphenous vein graft to the medial portion of the right coronary artery and the circumflex artery. Anatomically, the right ventricle was slightly enlarged, the complete left-sided pericardium was absent, and the heart was lying in the left

hemithorax with the apex almost touching the dorsal thoracic wall (Figure 2A). Positioning of the heart for the LAD bypass was accomplished by means of a combination of an Octopus and a Starfish stabilizer (Medtronic, Minneapolis, MN, USA) (Figure 2B). Access to the right coronary artery and the circumflex artery presented no relevant technical challenges attributable to the left-posterior rotation of the heart. Both the operative and postoperative courses were uneventful and the patient was discharged after 9 days.

COMMENT

Congenital absence of the pericardium is a rare entity that can occur in isolation or in association with other structural or genetical anomalies, including deformation of the thoracic organs [Van Son 1993; Dias 2007]. Because this condition in asymptomatic patients is usually discovered through postmortem or incidental diagnosis, it is difficult to ascertain its total prevalence, and the isolated form of this congenital defect comprises only a portion of the 0.0001% to 0.044% prevalence of the reported cases [Van Son 1993]. Embryologically, pericardial absence is believed to originate from the agenesis of the left common cardiac vein, which is the precursor of the left pleuropericardial membrane [Van Son 1993; Rashid 2008].

Congenital pericardium defects are usually asymptomatic, and the symptoms of patients with those defects are commonly heterogeneous. However, left-sided chest pain, which may be associated with changes in posture, is frequently the dominant symptom [Gatzoulis 2000]. Possible causes of pain in this condition include tension on pleuropericardial adhesions, ischemic-type pain due to compression of the coronary artery branches, or torsion and herniation of heart structures through the pericardial defect [Rashid 2008]. Patient symptoms are mainly related to signs of right-heart overloading as well as chest discomfort and angina. In the patient we present, however, the presence of multivessel artery disease may have masked the pericardial symptoms and vice versa. Not only the symptoms but also the clinical procedures are dependent on the intrathoracic displacement of the heart cavities. A few cases of severe tricuspid regurgitation and chronic right heart insufficiency due to the posterior left rotation of the heart have been described [Gatzoulis 2000; Goetz 2004]. This lateral displacement can also produce friction of the epicardium with the pericardial rim, causing local stenoses, which may derive from vascular lesions [Gersbach 1996]. In our patient, the presence of this pericardial defect was associated with right-heart overloading, probably due to the displacement, which caused a distortion of the left and right ventricular geometry. Furthermore, the localization of the coronary lesions, with medial stenosis in the LAD, circumflex, and right coronary arteries, was concordant with the pericardial defect.

A few cases reported in the literature about the simultaneous presence of congenital pericardium defects and coronary artery disease [Gersbach 1996; Nguyen 2001]. To our knowledge, the cases described until now were treated with cardiopulmonary bypass. However, as reported by Nguyen and colleagues [2001], cannulation for cardiopulmonary bypass in patients with congenital defects of the pericardium may

be associated with technical difficulties due to the clockwise rotation of the vascular structures. Furthermore, preoperative assessment of the graft material is mandatory. Both the arterial and the venous conduits must be longer than those used in patients with a normally positioned heart, because heart rotation makes the distances between the aorta and LITA to the left coronary system much longer. After considering the existing pericardial disease in our patient, we performed off-pump coronary bypass surgery for 2 reasons. This procedure enabled us to avoid cannulation difficulties that have been previously reported [Firstenberg 2006]. We also were able to assess the length of the grafts and the anatomical relationship of the LITA to the LAD without arresting the heart, and thus we avoided the “bowstring” phenomena after lung inflation or anastomotic disruptions. Our treatment proved effective, and we recommend such an approach in patients presenting with pericardial absence and associated coronary artery disease.

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