Incidental detection of a giant ductus arteriosus aneurysm by low-dose multidetector computed tomography in an asymptomatic adult

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Aneurysm of the ductus arteriosus is a very rare congenital lesion in adults that can be associated with thromboembolism, rupture, and death. Its detection in a silent clinical phase is very important for planning appropriate treatment and avoiding potentially fatal complications. We report a case of a patent ductus arteriosus aneurysm of very large size (65.5 mm) that was incidentally discovered with low-dose (3.2 mSv) multidetector computed tomography in an asymptomatic 67-year-old man. The presence of coronary disease was also ruled out with this non-invasive imaging modality. Further evaluation with echocardiography and selective angiography confirmed the diagnosis. At surgery, the aneurysm was successfully resected via a left posterolateral thoracotomy. (J Vasc Surg 2010;51:1260–4.)

Ductus arteriosus aneurysm (DAA) is a very rare entity in adults, particularly with evidence of communication between the aorta and the pulmonary artery.1 A large DAA may be recognized incidentally2 or may present with symptoms of a thoracic mass, most commonly in adulthood, and is associated with potentially life-threatening complications, such as rupture, erosion into bronchus or esophagus, infection, and thromboembolism.3,4 We report a case of a patent DAA of very large size in an asymptomatic 67-year-old man in whom the diagnosis was made by multidetector computed tomography (MDCT) using prospective electrocardiogram (ECG) gating. The presence of coronary artery disease was also ruled out with this non-invasive imaging modality. Successful surgical repair was performed via a left posterolateral thoracotomy without cardiopulmonary bypass.

CASE REPORT

A chest radiograph performed because of fever in a 67-year-old man with history of hypertension showed a rounded mass with faint calcifications in the aortopulmonary window (Fig 1). The patient was referred to our hospital for further evaluation of the chest mass. Physical examination revealed a grade 2/6 systolic murmur over the left upper sternal border. Laboratory studies and ECG were normal. To better define the lesion, chest computed tomographic scanning was performed. In consideration of the mass location in the aortopulmonary window close to the left ventricle profile, MDCT with prospective ECG gating was used with the aim of minimizing movement artifacts. This technique allowed also the reduction of effective radiation dose and the evaluation of the coronary arteries. A very large (65.5 mm) saccular DAA was identified (Fig 1). The aneurysm originated from the ventral part of the distal aortic arch and extended toward the left pulmonary artery that showed mild compression (Fig 1). Multiplanar reconstruction revealed DAA patency and ruled out significant coronary artery disease (Fig 2). The effective MDCT radiation dose was 3.2 mSv. Normal left and right ventricular systolic function, mild concentric left ventricular hypertrophy, and normal pulmonary artery pressure were demonstrated with echocardiography. A large cavity close to the left pulmonary artery was also observed in the parasternal short axis view, while two color Doppler flow signals were detected originating from the aortic arch and entering the origin of the left pulmonary artery, respectively (Fig 3). Aortic angiography was unable to show the DAA, whereas selective contrast injection clearly defined the aneurysm anatomy. Invasive coronary angiography confirmed the absence of coronary artery disease (Fig 3).

Due to the large dimension and unfavorable shape of the duct, we chose to perform a surgical ligation in alternative to transcatheter occlusion. A left posterolateral thoracotomy was performed. The thoracic aorta and the pulmonary artery were simultaneously clamped. The aneurysm was opened and the aorta and pulmonary artery orifices were closed. Clamping time was two minutes. The resected specimen showed a thin wall with several calcifications. Histologically, all three structural layers were represented, but...
severe degeneration was present with marked irregular fibrous thickening of the intima and focal disruption and thinning of the underlying muscle layer. The postoperative course was uneventful and the patient was discharged seven days later.

DISCUSSION

Aneurysmal dilatation of the ductus arteriosus is a congenital lesion more frequently found in children under two months of age.\(^5,6\) In most series, the reported incidence of DAA among fetuses and neonates ranged between 0.8% and 8%, while the maximal diameter of the aneurysm rarely exceeded 20 mm.\(^6,7\) Only a few cases of DAA in adulthood have been reported in the literature.\(^1,3,4,8,9\) Earlier reports included only patients in whom DAA was found incidentally during thoracotomy performed for other pathology\(^5,10,11\) or, like in our case, for an abnormal chest radiograph.\(^5,10,11\) Adult presentation of DAA is associated with higher mortality than in childhood.\(^12\) Indeed, life-threatening complications such as rupture, erosion, thromboembolism, and infection have been reported.\(^1,13\) The DAA of our patient was constantly subjected to systemic pressure due to direct communication with the aorta and, therefore, was at significant risk of rupturing. Several theories about DAA pathogenesis have been proposed, but the definitive mechanism of its formation remains uncertain. The common belief is that DAA may be the result of delayed closure of the aortic end of the ductus arteriosus with exposure of the ductal wall to systemic arterial pressure.\(^6,7\) Another potential mechanism of DAA formation is congenital weakening of the ductal wall due to reduced intimal cushion formation or abnormal elastin and extracellular matrix deposition.\(^6,7\) Indeed, the ductus closure process begins at the pulmonary artery end and progresses toward the aortic end, which then may not completely obliterate leading to an “aortic diverticulum.”\(^14\) It is noteworthy that in most of the ductus diverticulum aneurysms reported in the literature the aortic end was patent, whereas the pulmonary end was usually closed.\(^6,7\) Moreover, many adults with DAA are hypertensive, suggesting that progressive ductus dilatation may be favored by constant pressure...
overload. On the other end, an underlying congenital disorder that may lead to ductal wall weakening such as trisomy 21, trisomy 13, Smith-Lemli-Opitz syndrome, type IV Ehlers-Danlos syndrome, or Marfan’s syndrome is present in approximately one-fourth of patients with DAA. Most infants with DAA are asymptomatic. The adult form of this condition may become symptomatic relatively late, suggesting a slowly growing process. Presenting symptoms are those of a thoracic mass, including dyspnea, cough, chest pain, and hoarseness due to left vocal cord paralysis from recurrent left laryngeal nerve impingement. Symptoms secondary to pulmonary hypertension are rare in adults, since patency of DAA is very uncommon. A patent DAA of this size without symptoms at the age of our patient is unusual. The presence of the aneurysm was not suspected on previous medical examination, and the diagnostic work-up began with a chest radiograph showing a soft-tissue mediastinal mass-like shadow in the aortopulmonary window. The differential diagnosis included mediastinal tumor or abscess, lymph nodes enlargement, post-traumatic or atherosclerotic aneurysm of the thoracic aorta, aortic dilatation due to infection (ie, luetic aortitis), bronchogenic cyst, and DAA. Indeed, the diagnostic problems of plain chest radiography are related to the recognition of the vascular nature of the lesion. Parietal calcification at the periphery of the lesion and demonstration of a small pedicle linking the lesion to the pulmonary artery may suggest a DAA. However, large DAA that reduce and deform the mediastinal space may obscure the pedicle radiological sign.

The imaging protocol of patients with suspected DAA should include echocardiography after initial chest X-rays. However, a limited acoustic window in the adult patient may impair appropriate evaluation of this region and color Doppler, which is a very sensitive modality in detecting the presence of a patent ductus arteriosus, may be unable to detect a DAA when there is no left-to-right shunting. Alternative imaging techniques, such as magnetic reso-
nance or computed tomographic imaging, are ideal for showing the vascular nature of this lesion, because contrast enhancement and density similar to other vascular structures leave no doubts about the nature of the mass. In our patient, the diagnosis of DDA was made with MDCT using prospective ECG gating. The exact DAA dimension, wall characteristics, calcifications, and absence of thrombus were well seen with this imaging modality. Moreover, the exact relationship of the mass to adjacent mediastinal structures was precisely defined. Finally, with the same scan, DAA patency was identified and the presence of coronary artery disease was ruled out. It is noteworthy that all these imaging information were obtained with a lower effective radiation dose as compared to standard cardiac CT using retrospective ECG triggering and invasive angiography in which up to 20 mSv and 9 mSv, respectively, can be reached. Although it has been reported that angiography can be helpful to confirm the diagnosis of DAA, aortography failed to detect the lesion. This was likely due to the very small dimension of the pedicle linking the DAA to the aorta. However, the previous identification of the DAA with MDCT prompted the angiographer to search for it with selective catheterization that confirmed the diagnosis and the connections at the aortic and pulmonary ends.

Surgical ligation and resection remains the treatment of choice of DAA. The presence of extensive calcification in the wall of the aneurysm can hamper the opening of the sac, determining a longer clamping time. However, in our case, despite the presence of wall calcification, the total clamping time of the aorta and pulmonary artery was about two minutes. Transluminal endovascular stent grafting has been proposed as a feasible alternative based on the safety and efficacy demonstrated by this non-surgical approach for thoracic aortic aneurysm treatment. Saito et al reported successful endovascular repair of an aneurysm of the ductus diverticulum in a 78-year-old woman considered at high risk for surgery. An Inoue single-branched stent graft was used because the aneurysm was adjacent to the origin of the left subclavian artery. In our case, the surgical option was preferred. Indeed, our patient was not high risk for conventional open surgery. Moreover, given the DAA patency, endovascular repair with aortic stent grafting would have left a communication between the pulmonary artery and the aneurysm.

To the best of our knowledge, this is the first report of an adult asymptomatic patient in whom MDCT with prospective ECG gating and very low radiation dose identified a giant DAA, detected patency of the aneurysm defining the anatomy of the communications between the aorta and pulmonary artery, and ruled out the potential association with coronary artery disease.

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REFERENCES


