An Extremely Large Coronary Aneurysm Associated with a Quadricuspid Aortic Valve in an Adult Patient

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Abstract

A 68-year-old woman exhibited an increasingly protruding mass on the left heart border on chest X-ray. Transthoracic echocardiography revealed an echo-free mass in the anterior pericardial space. Transesophageal echocardiography revealed blood flow from the proximal left anterior descending coronary into a large coronary artery aneurysm measuring 61 mm × 51 mm in diameter and a quadricuspid aortic valve with a small cusp between the left and right coronary cusps. Coronary angiography demonstrated the presence of a coronary aneurysm connected to the proximal left coronary anterior descending artery. A giant coronary artery aneurysm and pulmonary artery fistulas extending from the left and right coronary arteries were confirmed by surgeons and successfully treated with surgery.

Key words: giant coronary aneurysm, coronary artery fistula, quadricuspid aortic valve, transesophageal echocardiography


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Introduction

A coronary artery aneurysm is defined as a coronary dilatation that exceeds the diameter of normal adjacent segments by a factor of 1.5 (1). The etiology of coronary aneurysms involves congenital malformation, atherosclerosis, chest trauma, Kawasaki disease, sarcoidosis, Behcet’s disease, systemic lupus erythematosus, directional coronary atherectomy and postimplantation of a drug-eluting stent (2-12). Coronary artery fistulas are a common congenital disease that is often combined with giant coronary aneurysms (13). A giant coronary aneurysm may cause coronary artery rupture, thromboembolism or hemodynamic problems related to compression. Rupture and cardiac tamponade may occur without the development of progressive dilatation (14). Therefore, making an earlier diagnosis is very important, and a noninvasive diagnosis may be useful for evaluating the time course of the disease and selecting a treatment strategy. We diagnosed the present patient noninvasively with an extremely large left coronary artery aneurysm measuring 61 mm in diameter in association with a quadricuspid aortic valve using echocardiography and multidetector row CT. We were not able to find this combination of anomalies reported in any recent literature and consider this diagnosis to be very rare.

Case Report

A 68-year-old woman with a history of treatment of hypertension presented to our outpatient clinic for examination of a protruding mass located at the left heart border on chest X-ray that had been increasing in size for three years. She felt no chest pain or exertional dyspnea. Her blood pressure was 144/85 mmHg and her heart rate was 70 beats/min. She had no cyanosis. A faint diastolic regurgitant murmur at the upper left sternal border was audible. On chest X-ray, the cardiothoracic ratio was 52% and the left upper heart border
was protruding; however, the pulmonary vasculature was not increased (Fig. 1). Electrocardiogram (ECG) showed a normal electrical axis and flat T waves in lead V_{5\text{a}}. A huge echolucent mass measuring 61 mm × 51 mm in diameter in the pericardial space was noted on transthoracic echocardiogram; however, the relationship of the mass to the left coronary artery was not well documented. Mild aortic regurgitation was also noted. The left ventricular diastolic dimension was 46 mm, the left atrial dimension was 42 mm and the left ventricular ejection fraction was 74%. On transesophageal echocardiography, the left main coronary artery measured 6 mm in diameter, the giant aneurysm was connected to the left anterior descending coronary artery and blood flow from the proximal left anterior descending artery into the aneurysm with a mural thrombus was observed (Fig. 2). The aortic valve was a quadricuspid aortic valve with three large cusps and one small cusp, with the small cusp located between the left and right coronary cusps on transesophageal echocardiography (Fig. 3). Multidetector row contrast-enhanced CT demonstrated the presence of a late enhanced mass measuring 50 mm × 62 mm in diameter (Fig. 4). The presence of a quadricuspid aortic valve was also confirmed on CT and contrast aortography (Fig. 4). Left coronary angiogram showed the jet flow from the proximal left anterior descending coronary artery (segment 6) into the huge aneurysm, while the flow in segment 7 of
Coronary artery aneurysms may be caused by coronary arteritis, congenital malformations, postcoronary interventions and chest trauma (2-12). The aneurysm in the present case was associated with coronary pulmonary artery fistulas and had a short trunk measuring 2-3 mm in length branching from the left anterior descending coronary. The presence of two exit vessels from the aneurysm to the pulmonary fistulas was confirmed during surgery. Therefore, the pathogenesis may be congenital and associated with coronary pulmonary artery fistulas. The size of the aneurysm measured on CT was very large (62 mm in diameter). Holinski reported the case of a 63-year-old man with two aneurysms of the circumflex coronary artery measuring 65 mm in diameter and noted that giant coronary artery aneurysms (defined as aneurysms larger than 20 mm in diameter) are extremely rare (15). The size of the aneurysm in the present case is the second largest reported in the literature. The prevalence of giant coronary aneurysms was reported to be six out of 30,268 patients who underwent heart surgery at Fuwai Hospital in Beijing (13). Five of these six cases were associated with coronary artery fistulas, as in our case. Making an earlier noninvasive diagnosis of coronary artery aneurysms may be clinically relevant because thromboembolism, rupture and hemodynamic problems related to compression may occur. Echocardiography and multidetector row CT are useful for diagnosing coronary aneurysms (3, 16, 17). In the present case, transthoracic echocardiography also visualized the giant echolucent mass in the pericardial space; however, transesophageal echocardiography was more useful for precisely visualizing the color Doppler flow into the aneurysm from the donor left anterior descending coronary artery. In addition, we were able to detect the quadricuspid aortic valve with mild aortic regurgitation on transesophageal echocardiography and multislice CT. Quadricuspid aortic valves are classified into five types: one small and three large cusps (67%), four equal cusps (18%), four unequal cusps (6%), three small and one large cusp (6%) and two small and two large cusps (3%) (18). Our present patient had one small and three large cusps with the small cusp noted between the left and right coronary cusps, the type that is most frequently observed in the literature. There is one case report of two aneurysms associated with coronary artery fistulas and a quadricuspid aortic valve.
The authors state that they have no Conflict of Interest (COI).

References


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