CASE REPORT

Coronary artery aneurysm with a fistulous connection to the right atrium mimicking a sinus of Valsalva aneurysm

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Coronary artery aneurysms are uncommon and may be complicated by rupture, thromboembolic phenomenon, and more rarely fistulation into one of the cardiac chambers. This case report highlights the difficulty in making a preoperative diagnosis of a coronary artery aneurysm that has fistulated into the right atrium, and lists possible differential diagnoses.

The incidence of coronary artery aneurysms in patients undergoing coronary artery angiography is 1.5–4.9%. They occur most commonly in the proximal right coronary artery (RCA) and may be complicated by rupture and thromboembolic phenomenon.^{1,2} In this case there was a fistulous connection between an aneurysmal proximal RCA and the right atrium, which was initially diagnosed as a sinus of Valsalva aneurysm by transoesophageal echocardiography (TOE) and subsequently confirmed by coronary angiography. At surgery, the patient was found to have a large RCA aneurysm with a fistulous connection to the right atrium without any evidence of associated cardiac abnormalities. This case illustrates the difficulty of locating the proximal end of a coronary artery to cardiac chamber fistula radiographically.

CASE PRESENTATION

A 43 year old woman presented with increasing dyspnoea on exertion. On examination, the only abnormality detected was a soft left parasternal systolic murmur. Chest radiography showed cardiomegaly with pulmonary plethora. A sinus of Valsalva aneurysm was clinically diagnosed. TOE (video sequence 1) showed a sinus of Valsalva aneurysm that communicated with the right atrium and this was later confirmed by coronary angiography (video sequence 2), although the possibility of a fistula from the RCA to the right atrium was considered.

At operation, the aortic valve and the sinuses of Valsalva were normal. The origin of the RCA was aneurysmal with a diameter of 1.5 cm over its proximal 3 cm with a fistulous connection to the right atrium. The right atrium was opened, exposing the entrance of the fistula located just superior to the border of the fossa ovalis. The extracardiac portion of the fistula was then excised and the defect in the RCA closed in two layers. The defects in the right atrium and the area where the fistula had emerged from the myocardium were also closed in two layers. An interposition vein graft was not used because the RCA distal to the aneurysm was very small and anastomosis to a graft was not feasible. In view of the dominant left system, the RCA distal to the fistula was ligated.

DISCUSSION

The most common cause of coronary artery aneurysms is atherosclerosis followed by Kawasaki disease, periarteritis nodosa, systemic lupus erythematosus, syphilis, rheumatic fever, congenital heart disease, and trauma.³ Fistulous connection to the cardiac chambers may result from rupture of a coronary artery aneurysm or it may be the primary anomaly that is then complicated by a coronary artery aneurysm.

The indication for surgery on coronary artery aneurysms depends on the size and the risk of rupture, the presence of concomitant obstructive coronary artery disease, and the presence and size of a fistula to one of the cardiac chambers.⁴ In this case, even though the origin of the fistula was not confirmed, the presence of symptoms and signs of a left to right shunt and the presence of a fistula to the right atrium justified surgery. The differential diagnosis of an RCA to atrial fistula includes the rupture of a sinus of Valsalva aneurysm, a coronary arteriovenous fistula, and rupture of a dissecting aneurysm of the ascending aorta into the right atrium.⁵

TOE with colour flow Doppler gives good views of the proximal coronary arteries from the oesophageal window and may be comparable with the ideal of coronary angiography.^{6,7} We present this case to highlight the difficulty in diagnosing coronary artery to cardiac chamber fistulas despite using TOE and coronary angiography.

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