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Images in cardiology

Aneurysm of the right sinus of Valsalva and the membranous septum: Haemodynamic and clinical characterization

Ahmed Afifi*, Sherine AbdelSalam, Ahmed ElGuindy, Mohamed Donya

Aswan Heart Centre, Aswan, Egypt
*Email: ahmedafifi@doctors.org.uk**INTRODUCTION**

Sinus of Valsalva aneurysms (SVAs) constitute about 1% of congenital cardiac anomalies and occur most commonly in patients of Asian origin.¹ It is most commonly described in the right sinus (70%), to a lesser degree in the non-coronary, and least common in the left sinus. These aneurysms can cause aortic regurgitation, cardiac ischemia and arrhythmias. They can rupture into the right cardiac chambers or the interventricular septum. We here describe an aneurysm of the right coronary sinus with unique clinical and haemodynamic features.

CASE REPORT

A 26-year-old male patient was admitted with acute onset of dyspnoea, orthopnoea and haemoptysis. On admission his blood pressure was 160/80 and his ECG showed sinus bradycardia 60/min. Echocardiography showed severe aortic regurgitation, moderate mitral regurgitation, and a “peculiar” morphology of the aortic root. The right sinus of Valsalva was enlarged (4 × 6 cm), protruding into the interventricular septum and encroaching on the right atrial cavity. The aneurysm, in addition to bulging into the right ventricle in diastole, was also pushing the membranous septum, making it aneurysmal (Figure 1). The right sinus was communicating with the membranous septum via two perforations, which produced a bilobed bulge into the left ventricular outflow tract (Figure 2a and c). This marked distortion of the valve morphology caused severe aortic incompetence. In systole, the bulge regressed, leaving the outflow tract wide open (Figure 2b and d). A CT was performed (Figure 3) showing an aneurysm of both the right sinus of Valsalva, dissecting into the interventricular septum causing aneurysm formation of the membranous interventricular septum that bulged into the right ventricle, and another in the atrio-ventricular septum, with a bulge into the right atrium.

The distinctive findings on imaging confirmed the diagnosis of an un-ruptured right sinus of Valsalva aneurysm. The patient underwent urgent surgery of the aortic root. A Bentall procedure was performed with size 29 mechanical valve, 30 mm ascending aortic Dacron tube graft, and re-implantation of the coronary buttons. Intra-operative findings were in agreement with the echocardiographic data (Figure 1–left). The dissected flap was excised (Figure 1–right) and pledgeted, mattress sutures were taken from the healthy part of the septum, distal to the aneurysm, and these sutures were passed through the aortic annulus and used to secure the aortic prosthesis in position. Sutures were taken in the annulus as usual in the left and noncoronary position. The operation was complicated by complete heart block that required permanent pacing. On follow-up echocardiography the membranous septum, which weakened by the chronic aneurysm, developed a pseudo-aneurysm in close proximity to the aortic prosthesis which was managed percutaneously three weeks later.

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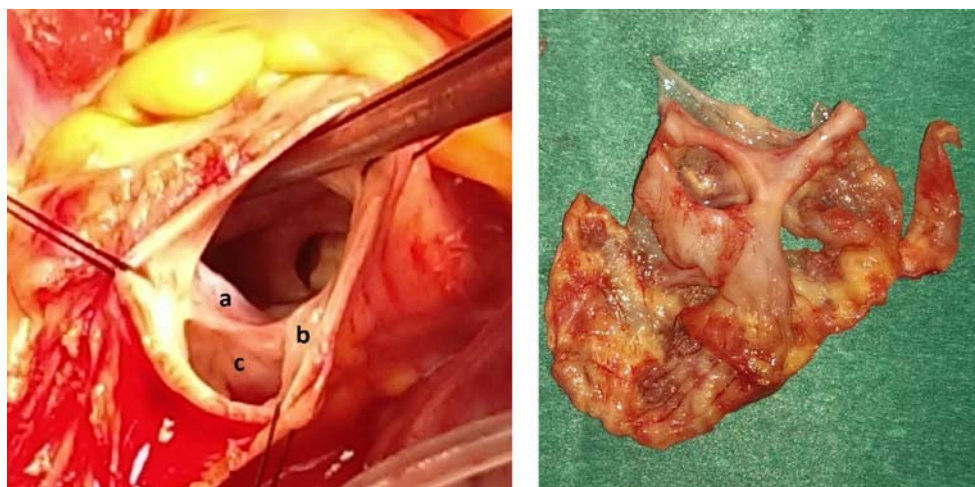


Figure 1. Surgical view – Left: Surgical view of the aortic root showing the large right sinus of Valvsalva aneurysm anteriorly and the three aortic cusps (a: RCC, b: NCC, c: LCC) Right: Surgical specimen of the dissected interventricular septum with the RCC attached at the top.

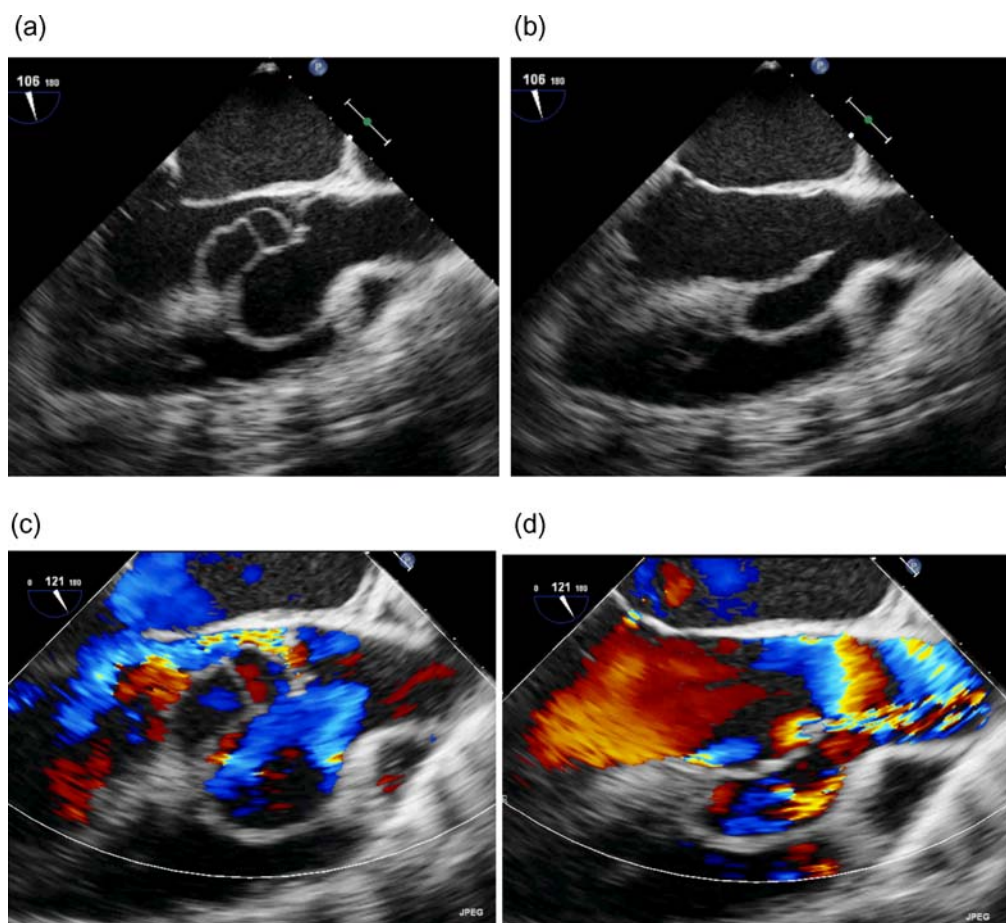


Figure 2. Transesophageal echocardiography (Long axis view) *a and c*: the aortic root in diastole, the sinus aneurysm fills and bulges into the right ventricle, the membranous septum is aneurysmal and obliterates the left ventricular outflow tract, severe eccentric aortic incompetence is shown and there is a communication between the right sinus and membranous septum aneurysm. *b and d*: the aortic root in systole, the sinus aneurysm empties and the bulge in the right ventricle is reduced, the membranous septum aneurysm collapses and the LVOT is wide open.

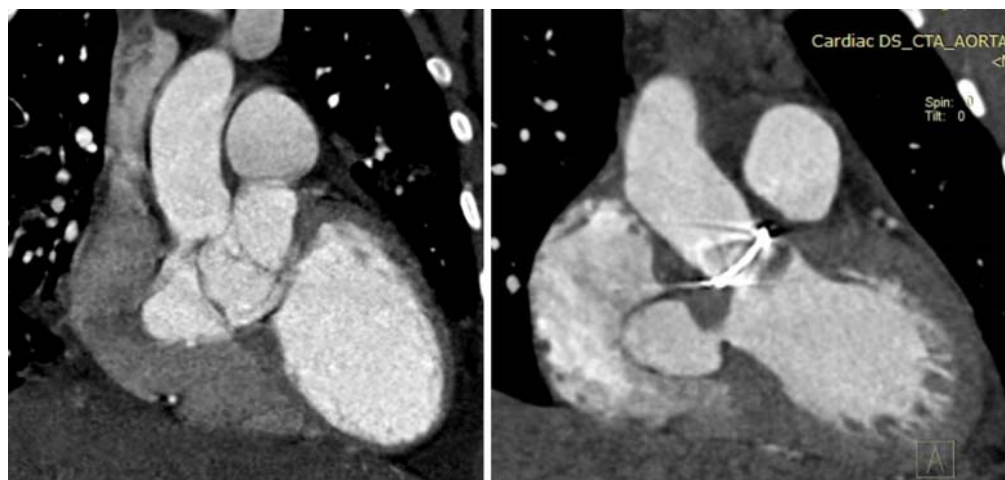


Figure 3. Multi-slice computed tomography of the aortic root *left*: preoperative: dilated sinus of valsalva aneurysm with extension into the septum, the membranous septum is bulging into the left ventricular outflow *right*: postoperative: the aneurysm is obliterated, the aortic prosthesis is in place and a false aneurysm of the membranous septum is seen encroaching on the right atrial cavity.

DISCUSSION

Congenitally, sinus of Valsalva aneurysms are thought to be due to failure of fusion between the media of the aortic wall and the annulus fibrosus of the aortic valve. They can also be acquired, as a result of injury, endocarditis, syphilis or Behcet's disease.² SVAs may rupture in the 3rd or 4th decade of life, usually in the right atrium or right ventricle, into the interventricular septum or, rarely, in the pericardial cavity.³ Ruptured SVAs can cause progressive heart failure, caused by the left to right shunt and/or aortic regurgitation, or complete atrioventricular block caused by interventricular septal rupture. Unruptured SVAs can cause thrombus formation, with subsequent embolic events, myocardial ischemia, due to compression of a coronary artery by the body of the aneurysm, or obstruction of the right ventricular outflow.⁴

Echocardiography is the diagnostic method of choice and transesophageal echocardiography is especially important to guide surgical management. It provides important information on the site, size and morphology of the sinus aneurysm. It also detects coexisting lesions, complications and haemodynamic sequelae.⁵ Computed tomography can be of added value for precise determination of size, extent and anatomical relations. Surgical repair is indicated if the size of the aneurysmal sinus exceeds double that of the healthy ones, or if complications start to arise - although surgical intervention is encouraged before the onset of complications, since the perioperative complication rate is comparatively very low. A number of surgical and transcatheter approaches have been described like direct suturing or patch closure of the "mouth"/inlet of the aneurysm and closure of the exit of the sinus rupture through the right atrium or ventricle.^{2,6}

LESSONS LEARNED

Defining the exact haemodynamics and sites of extension or rupture of the aneurysm can be useful for identifying if extra procedures are needed in addition to closing the mouth. In this case we believe that the weakened membranous septum was the cause of the false aneurysm discovered during follow up. It would have been justified to inspect the membranous septum intra-operatively for the possibility of strengthening/supporting it without encroaching on the conduction tissue.

Given the severe dilatation and abnormal anatomy of the aortic root, replacement with a Bentall procedure was considered a safer option. However, the preferred approach would be closing the opening/entry of such aneurysms from within the aorta. In that way the aneurysm will cease to become a high pressure chamber and subsequently the aneurysm cavity will regress and obliterate. Early surgery, before the onset of severe symptoms or complications, may also allow for a lower risk reconstructive surgery with the benefit of a valve sparing operation.

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