Giant ruptured sinus of Valsalva aneurysm: diagnosis on echocardiography

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A giant ruptured sinus of Valsalva aneurysm was diagnosed on transthoracic and subsequent transesophageal echocardiography, in a 45-year-old man who presented with gradual onset shortness of breath. Although the initial presentation was insidious, he later rapidly deteriorated. We discuss the unusual clinical course in a patient with such a large aneurysm and discuss the likely reasons.

KEYWORDS
Congenital heart disease; Rupture; Aneurysm

A 45-year-old man presented with a three-month history of atypical chest pain and exertional dyspnea. He was noted to have a grade four continuous murmur, loudest over the right 4th intercostal space. A chest X-ray showed cardiomegaly, widening of the mediastinum and pulmonary plethora. Transthoracic echocardiography appeared to show a very large aneurysm of the non-coronary sinus of Valsalva, which protruded into the right atrial cavity. (Figure 1/Movie clip 1). There was a small rupture of the aneurysm into the right atrium. These findings were clarified on transesophageal echocardiography (Figure 2, Movie clips 2 and 3). Cardiac catheterization was performed, which showed a step-up in oxygen saturations from 76% (mixed venous) to 90% in the right atrium (left-to-right shunt of 2.4:1) and an elevated right atrial pressure of 20 mmHg. There was mild pulmonary arterial hypertension (mean pulmonary arterial pressure 27 mmHg). Angiography further demonstrated a giant aneurysm of the non-coronary sinus of Valsalva (Figure 3, Movie clip 3).

Unfortunately the patient developed a chest infection and so his operation was postponed for a few days. He then suddenly went into gross right sided heart failure, although repeat echocardiography failed to demonstrate worsening of the rupture. He needed stabilization with fluid, inotropes and cautious use of diuretics. Although he became haemodynamically stable once again, the risk of operation had considerably increased, with echocardiography showing severe right ventricular dysfunction. After consultation with the cardiologists and surgeons, the patient opted for medical therapy alone and was discharged to the care of his local physician.

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Aneurysms of the sinuses of Valsalva comprise about 1% of congenital heart disease. They most commonly arise from the right coronary sinus (65–85%), also from the non-coronary sinus (10–30%) and rarely the left coronary sinus (<5%). The primary concern is of rupture and this is most commonly into the right ventricle (90%), occasionally into the right atrium (10%) and rarely into the left heart chambers or pericardium. However, the aneurysm and rupture are usually small and take on the appearance of a ‘windsock.’ Previous cases of giant aneurysms which have ruptured have required immediate surgical intervention. Our case demonstrates that even with a giant aneurysm of one of the sinuses of Valsalva, the clinical course of rupture may be insidious and survival may be possible without emergency intervention. However, the further developments in this case exhibit the importance of not delaying surgery once a diagnosis has been made, due to the risk of developing further complications.

Echocardiography, particularly transesophageal, appears to be the imaging modality of choice in detailing the anatomy of this rare condition.

Supplementary material
Supplementary material associated with this article can be found in the online version.

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References