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Brief Report

Sinus of Valsalva aneurysm rupture in an infant

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Abstract We present the case of a 10-month-old female with a right coronary sinus of Valsalva aneurysm with rupture into the right atrial appendage who presented with a murmur. Surgical repair was performed shortly after diagnosis with pericardial patch closure from within the aorta and closure of the right atrial rupture site. To our knowledge, this is the youngest child with sinus of Valsalva aneurysm with rupture to be identified in the literature.

Keywords: CHD; sinus of Valsalva aneurysm; infant

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For paediatric cardiologists and cardiothoracic surgeons alike, cases of sinus of Valsalva aneurysms with rupture are rare entities. They account for 0.5–3% of all CHD.[1] These defects are uncommonly detected in the paediatric age range because the median age of rupture, and subsequent murmur that brings most patients to the attention of a cardiologist, is 31 years.[2] We highlight this case of a 10-month-old infant with ruptured sinus of Valsalva aneurysm to ensure it is on the differential of paediatric cardiologists when confronted with an infant with a new-onset murmur, signs of heart failure, or with echocardiographic findings of intracardiac left-to-right shunts or aortic insufficiency.

Case report

A 10-month-old full-term female with no significant past medical history was referred to an outside paediatric cardiologist for a history of an abnormal "mediastinal shadow" on prenatal ultrasound scanning. Physical examination at that time revealed a continuous murmur and the cardiologist felt that the echocardiogram was concerning for right coronary artery to main pulmonary artery fistula. The patient was then referred to our institution for further evaluation.

On presentation to us, she was completely asymptomatic. Her mother denied any difficulty with feeding, lethargy, sweating, or loss of consciousness. She had been gaining weight appropriately and meeting all developmental milestones. The examination was only notable for a II/VI continuous murmur, heard best at the right upper sternal border. The rest of her examination was unremarkable. Her blood pressure was 101/57 mmHg and pulse was 127 bpm. Her weight was 8 kg, which was just above the 30th percentile. Electrocardiogram showed normal sinus rhythm with no ST elevation, heart block, or tachycardia.

Echocardiography on admission showed a ruptured right aortic sinus of Valsalva aneurysm (Fig 1a). The aneurysm was oblong, had a proximal dimension of 4.6 mm and a distal one of 3.1 mm on echocardiography, and was at a length of 13 mm. A 1.5 mm perforation was noted at the distal end with flow entering the right atrium and a systolic pressure gradient of 64 mmHg. The right coronary artery arose separately from the aneurysmal right sinus and appeared normal in size (Fig 1b).

A computed tomographic angiogram was obtained, which confirmed the echocardiographic findings (Fig 1c). Specifically, the right coronary artery originated separately from the sinus of Valsalva aneurysm and was normal in appearance.

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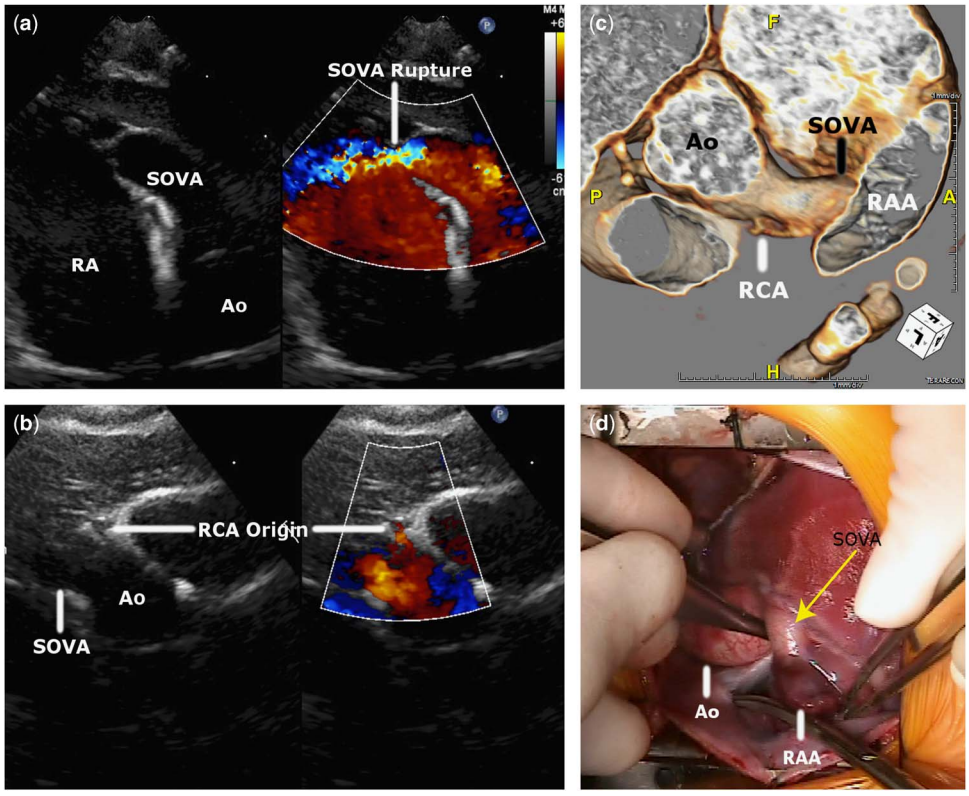

Figure 1.
*(**a**) Transthoracic echocardiogram: parasternal short axis view of SOVA and flow through the communication to the RA. (**b**) Transthoracic echocardiogram: parasternal short axis view of SOVA and separate origin of the RCA. (**c**) Computed tomographic angiography 3D reconstruction of SOVA. (**d**) Gross anatomic findings in the operating room. Ao = aorta; RA = right atrium; RAA = right atrial appendage; RCA = right coronary artery; SOVA = sinus of valsalva aneurysm.*

The right atrium was mildly dilated, but there was no evidence of right ventricle dilatation.

The patient was admitted to the step-down unit for anticipated surgery; however, the surgery was delayed as the patient had evidence of a viral illness with respiratory symptoms. On day 7 at the hospital she was sent to the operating room to undergo repair. In the operating room, a large sinus of Valsalva aneurysm, in very close proximity to but posterior and rightward of the right coronary artery ostium, was observed. The aneurysm had ruptured into the base of the right atrial appendage (Fig 1d). The sinus of Valsalva aneurysm opening into the aorta was roughly 6–8 mm in diameter and 15–20 mm long. Pledgeted sutures were used to close the right atrial end of the aneurysm and an autologous pericardial patch was used to close the aortic end of the defect. The patient came off-pump without difficulty, total bypass time being 95 minutes, and a postoperative transoesophageal echocardiogram showed no evidence of residual aneurysm or shunts. She was then transferred to the paediatric cardiac ICU in stable condition. She had an unremarkable postoperative course; specifically, she was extubated on postoperative day 0, removed off all vasoactive drips, and was put on room air and on the step-down unit by post-operative day 2. She was discharged home on post-operative day 5.

Discussion

To our knowledge, this is the youngest case of a ruptured sinus of Valsalva aneurysm documented in the literature. It should be included in the differential diagnosis of patients with continuous murmur. In order to differentiate this lesion from a right coronary artery fistula, it was important to identify the right coronary origin from the sinus of Valsalva separately from the origin of the aneurysm.

If detected early, this defect can be treated surgically or in the catheterisation laboratory before overt signs of heart failure develop. It is important to note, however, that a sinus of Valsalva aneurysm can cause clinical issues even if it does not rupture. For example, if large enough, the aneurysm can obstruct the right ventricular outflow tract. It may also distort the aortic valve causing aortic insufficiency; it can compress the left coronary artery causing signs of ischaemia; and, finally, it may compress the conduction system causing conduction delays or even complete heart block.[3]

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Two-thirds of sinus of Valsalva aneurysms arise from the right aortic sinus: as seen in this patient, one-fourth in the noncoronary sinus, and the remaining in the left aortic sinus.[2] A congenital lack of continuity between the aortic media and annulus fibrosis may predispose a patient to aneurysm formation.[4] The defect may also be acquired, being associated with the pathophysiology underlying mixed connective tissue disorders, endocarditis, syphilis, and trauma.

Surgical repair of a ruptured sinus of Valsalva aneurysm has excellent outcomes, with minimal risk for recurrent aneurysm formation and a small risk for aortic insufficiency later in life.[5] This further warrants the utility of early diagnosis to prevent the negative sequelae of large aneurysms with or without rupture.

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Conflicts of Interest

None.

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