Case report - Aortic and aneurysmal
Adult aortic coarctation discovered incidentally after the rupture of sinus of Valsalva aneurysm: combined surgical and interventional approach

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Abstract
Combination of ruptured sinus of Valsalva aneurysm (SVA), and a coexisting asymptomatic adult aortic isthmic coarctation is extremely rare. The timing and sequence of surgical and/or interventional repair of these two pathologies are controversial. We present a case of a 37-year-old male who was admitted to our department because of severe acute congestive heart failure and signs of ruptured aneurysm of the SV into the right ventricle. Transthoracic and transoesophageal echocardiography confirmed the communication between an important right coronary SVA and right ventricle, bicuspid aortic valve, mild aortic regurgitation, and revealed severe aortic coarctation. Because of the severe dilation of right sinus of Valsalva a surgical repair of the ruptured aneurysm was performed. Aortic coarctation was treated four weeks later by a percutaneous stent-graft implantation. This case report supports the concept that hybrid approach is feasible in patients with ruptured SVA and aortic coarctation in adulthood.

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1. Introduction
Congenital sinus of Valsalva aneurysm (SVA) is a rare cardiovascular anomaly and its rupture into right ventricle is further a rare occurrence [1–3]. The association of ruptured sinus of Valsalva to bicuspid aortic valve and aortic isthmic coarctation is exceptional. We describe here, a patient with all of these three lesions, who was successfully treated in a two-stage approach. Previously published reports of the coexistence of ruptured SVA and aortic coarctation, and their management, are discussed herein.

2. Case report
A 37-year-old male, with no previous medical history or cardiovascular risk factors, was admitted for abrupt onset of dyspnea (New York Heart Association III) over the previous two weeks. On examination, he was apyrexial with a blood pressure of 170/60 mmHg, a heart rate of 85 bpm and signs of right heart failure. A continuous murmur was audible at the second intercostal space on the sterna border. There were rales at both lung bases. The radial and femoral pulses were normal. No pretibial oedema was observed, nor were there any remarkable laboratory findings. Chest radiography showed cardiac enlargement (cardiothoracic ratio, 58%) with mild congestion of the pulmonary vasculature. Electrocardiography indicated sinus rhythm without any sign of hypertrophy. Cardiac transthoracic and transoesophageal echocardiography (Hewlett-Packard 5500 Sonos, Andover, MA, USA) showed a hugely dilated (51 mm) right sinus of Valsalva, a normal aortic root size (35 mm), bicuspid aortic valve, mild aortic insufficiency and a 9-mm communication between right coronary sinus of Valsalva and right ventricle. A severe aortic isthmic coarctation was also documented. Left ventricular (LV) dimensions and ejection fraction (63%) were normal. No evidence of infective endocarditis or coexistence of associated subaortic membranes was identified. Doppler analysis highlighted a left-to-right high-speed shunt flow from the right coronary SVA to the right ventricle.
Chest computed tomography (CT)-scan confirmed the SVA and the aortic isthmic coarctation with bilateral collateral arteries (Fig. 1). Heart catheterisation was not performed as it was felt it would not have given further information useful in the management of this patient.
Because the transcatheter closure of ruptured SVA was not available at that time and the SVA was particularly important (51 mm of diameter), the patient underwent surgical repair on cardiopulmonary bypass. The initial surgical
view showed right coronary SVA. Macroscopic analysis after transversal aortotomy confirmed the ruptured aneurysm of the right sinus of Valsalva in the right ventricle and the bicuspid aortic valve. The diameter of the communication was 10 mm. The openings of the aorta and the right ventricle were directly sutured with reconstruction of the anterior Valsalva sinus. Histological and microbiological analyses were negative. The patient made a good recovery except for high level of blood pressure, and was prescribed an angiotensin-converting enzyme inhibitor and calcium channel blockers at hospital discharge.

Four weeks after cardiac surgery, the patient was readmitted for management of the aortic coarctation by percutaneous stent implantation (Fig. 2). Percutaneous entry was initially made into the femoral artery for angiography showing a complete occlusion of the coarctation site (Fig. 2, Panel a). The guide wire could not be advanced through the aortic coarctation. Angiography in the ascending aorta across the radial artery showed a severe aortic coarctation (Fig. 2, Panel b). With the aid of a guide catheter, a long guide wire introduced into the radial artery was placed across the stenotic area. The guide wire was then advanced to the descending aorta where its proximal tip was snared and pulled through the femoral artery. A long sheath introduced into the femoral artery was then advanced across the lesion over the guide wire. The tip of the wire was positioned in the ascending aorta. The dilator was then removed and a balloon expandable stent was advanced inside the sheath to the coarctation site. Care was taken not to fully expand the balloon in order to reduce the likelihood of aortic wall damage. Postdilatation aortography showed no residual gradients (Fig. 2, Panel c). The procedure was uncomplicated and the patient was released the following day with antihypertensive medications. Control echocardiography (ECG) showed no communication with regression of the isthmic gradient. At follow-up two months postoperatively the
patient was asymptomatic. Repeat CT-scanning showed no recoarctation or aortic aneurysm formation.

3. Discussion

Coexisting lesions are common in patients with congenital, ruptured SVA [1–3]. Subarterial and perimembranous ventricular septal defects occur in Western patients with an incidence of 30–50%. Aortic regurgitation in ruptured SVA is also a commonly associated lesion, with 33.6% in the Asian group and 32.7% in the Western group. Bicuspid aortic valve occur with an incidence of 10–20% [2, 3]. Coexisting aortic coarctation was documented in 4% (3/86) of SVAs [1].

The association of ruptured SVA to aortic coarctation is exceptional and reported at a rate of only 1/57 of ruptured SVA [2]. Only few cases have been described so far [2, 4–6]. Over a cohort of 580 corrected cases of aortic coarctation, Manganas et al. [5] have reported that 23 patients required operation after coarctation repair. Only one patient have been re-operated for a ruptured SVA (1/580 patients with aortic coarctation). In our case report, the aortic coarctation and bicuspid aorta were discovered by chance at adulthood after the rupture of the SVA.

For the correction of combined forms of aortic coarctation and SVA with or without rupture, different techniques have been employed. In the one-stage repair, simultaneous correction of both lesions through a median sternotomy was performed [7, 8]. The two-stage repair can be performed through a combination of lateral thoracotomy and median sternotomy [4, 9]. Hybrid approach was also employed with the combination of the interventional and the surgical approaches [10]. The non-surgical treatment of the aortic coarctation proved to offer a significant advantage, converting a complex and risky surgical procedure into one of common practice [10].

Surgery used to be the mainstay of treatment of ruptured SVA, however, in past few years several reports of transcatheter closure of ruptured SVA have come to light [6]. Interventional treatment can be proposed to treat the two lesions with a device closure with a duct occluder device for the ruptured of SVA and percutaneous stent implantation for the aortic coarctation. Recently, Kerkar et al. [6] have reported the mid-term results of transcatheter closure using the first-generation Amplatzer duct occluder in patients with ruptured SVA who have no associated ventricular septal defect or aortic regurgitation. One patient had co-existing aortic coarctation. The rate of the procedure success was 90% with encouraging short- and mid-term outcomes.

4. Conclusion

Association of ruptured SVA to aortic coarctation is possible. Management of this complex association should be discussed case by case. Hybrid approach or totally interventional approach can be effective and safe in selected patients.

References