Case Reports

Simultaneous transcatheter closure of ruptured sinus of Valsalva aneurysm and stent implantation for aortic coarctation

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Ruptured sinus of Valsalva aneurysm is a rare anomaly and an associated coarctation of aorta is even rarer. A combination of such defects is traditionally treated surgically. The surgery is necessarily staged and done through different approaches. We report successful simultaneous transcatheter treatment of both these defects performed in the same setting in an acutely ill adult male patient with a good intermediate-term follow-up.

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1. Introduction

Ruptured sinus of Valsalva aneurysm (RSVA) is a rare, though well-recognized, clinical entity. It usually presents acutely or subacutely in adolescence to early adulthood. The aneurysm is usually congenital in origin and ruptures most commonly into a right-sided heart chamber. The association of RSVA with coarctation of aorta (CoA) is extremely rare.1 Stent implantation for CoA in adults is considered to be an acceptable alternative to surgery.2 In recent times, transcatheter closure (TCC) of RSVA is being proposed as a promising alternative to surgery in suitable patients.3 We describe here interventional therapy by the transcatheter approach for CoA and RSVA in the same setting as in an acutely ill patient.

2. Case report

A 32-year-old male presented with rapidly progressive dyspnea over a period of 10 days (New York Heart Association (NYHA) Class III). On physical examination, he was in congestive heart failure (CHF) with tachycardia, raised jugular venous pressure with prominent “V” waves, and bounding brachial and weak femoral pulses. The blood pressure (BP) in both arms was 160/50 mm Hg. Auscultation revealed fine rales over lung bases and a grade IV/V continuous murmur in lower left parasternal area. The electrocardiogram showed left ventricular hypertrophy and radiography was suggestive of cardiomegaly with increased pulmonary vascularity and rib notching. Transthoracic echocardiography (TTE) followed by
intraoperative transesophageal echocardiography (TEE) confirmed the presence of RSVA arising from noncoronary sinus (NCS) and draining into the right atrium (RA) resulting in a large left to right shunt, dilatation of all 4 chambers, and mild grade I/IV aortic regurgitation (AR) with normal biventricular systolic function. The RSVA measured 11 mm at the aortic end and 6 mm at the RA exit. Suprasternal view showed severe postsubclavian CoA with trivial anastomosis flow. After stabilization for 3 days, patient was taken up for cardiac catheterization under general anesthesia with TEE guidance. Before procedure, an informed consent was obtained and injection Cefazolin was administered. Right femoral venous and arterial accesses, and a right radial access, were established. Intravenous Heparin 100 U/kg was given. Cardiac catheterization demonstrated elevated right heart pressures (right atrial mean 14 mm Hg, right ventricle 40/16 mm Hg, and mean pulmonary artery pressure 30 mm Hg), elevated left ventricular (150/20 mm Hg), and ascending aortic pressure (150/60 mm Hg). The descending aortic pressure was 120/55 mm Hg, with a peak systolic gradient across the CoA of 30 mm Hg. The CoA was crossed with the help of 5F Judkins right (JR) coronary artery diagnostic catheter over a 0.035" exchange length straight-tipped glide wire and the catheter was exchanged with a pigtail catheter for diagnostic angiography in the descending aorta above the CoA (Fig. 1A, Video 1) and in the aortic root (Fig. 2A, Video 3). There was severe discrete CoA beyond left subclavian artery with the isthmus and the descending aorta at the level of diaphragm measuring 14 mm and 18 mm, respectively. Aortic root angiography confirmed RSVA arising from NCS and draining into the RA and mild aortic regurgitation (AR). A 39 mm long Cheatham-Platinum (CP) stent (NuMed, Hopekinton, NY) was hand-crimped on a 14 mm × 4 mm and 4 cm Z-MEDII (NuMed, Hopekinton, NY) high pressure balloon and passed through a long 12F sheath (Cook Inc., Bloomington, IN) over a 0.035 in. extra-stiff guidewire positioned in the ascending aorta. The stent was then precisely deployed at CoA site at 4 atm pressure under angiographic control. The stent was postdilated with a larger diameter (18 mm) balloon in the poststenotic segment for better conformation and a good result was confirmed with angiography. Poststenting, there were no gradients across the CoA taken with a multitrack catheter over the guidewire.

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The patient tolerated the procedure well and then we proceeded to perform TCC of RSVA. The RSVA was crossed from the aortic side using a 5F JR catheter over a 0.035 in. angled tip glide wire (Terumo Inc., Japan). This was exchanged for a 300 cm long noodle wire (St. Jude medical, Golden Valley, MN) that was snared with a 15 mm Amplatz gooseneck snare (ev3 Europe, Paris, France) from the superior vena cava (Fig. 2B, Video 4) and exteriorized out of the femoral vein. A stable arteriovenous wire loop was thus established, over which an 8F Amplatz delivery sheath (St Jude Medical, St. Paul, MN) was placed in the ascending aorta. An initial attempt at placing the largest available (16 mm × 14 mm) Amplatz Duct Occluder (St Jude Medical, St. Paul, MN) failed, as the device slipped through the defect into the RA. Although we could have used the radial route, we re-crossed the RSVA from the femoral route, carefully manipulating the catheter and guidewire across the stented CoA segment and reestablishing the arteriovenous wire loop. Under TEE, and fluoroscopic and angiographic guidance, we were able to position a larger 18 mm × 16 mm Lifetech Duct occluder (Shenzhen Lifetech Scientifc Inc., China) through a 9F SteerEase sheath (Shenzhen Lifetech Scientifc Inc., China) precisely across the defect (Fig. 2C, Video 5) taking care to avoid encroachment on the aortic valve and coronary arteries. After confirming no increase in AR or any residual shunt on TEE, the device was released by unscrewing the cable. A postprocedure angiogram in the aortic arch done through radial route confirmed optimal device position with complete occlusion of the defect and no increase in the grade of AR (Fig. 2D, Video 6) and good result across the stented coarctation segment (Fig. 1B, Video 2).

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The total procedural and fluoroscopy times were 125 min and 36 min, respectively. In the immediate postprocedure

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**Fig. 1 – A.** Descending thoracic aortogram in shallow left oblique caudal projection showing discrete severe postsubclavian coarctation of aorta with a poststenotic dilatation and B. Arch angiogram in the same projection showing relief of the coarctation with a precisely deployed Cheatham-Platinum stent.
3. Discussion

Ruptured sinus of Valsalva aneurysm is a very rare entity. Although coexisting lesions like ventricular septal defect and AR are commonly described with RSVA, the association of RSVA and CoA is very rare. In a western series, coexisting CoA was documented in 4% (1/86) while another Asian series reported a rate of 1 out of 57 RSVA cases. Traditionally, a combination of RSVA and CoA has been treated surgically. However, recently, a combination of RSVA and incidentally detected CoA were dealt with by surgical correction of RSVA followed by stenting for a CoA four weeks later. We have a fairly long and large experience of TCC of RSVA including interventional treatment of coexisting defects. Hence we decided to offer interventional treatment of this rare combination of defects. This case report is unique that both RSVA and CoA were treated nonsurgically with a single procedure in an acutely ill patient.

Unruptured aneurysms can remain undetected till they rupture into one of the chambers of the heart. Also, patients with isolated CoA may be incidentally detected due to high arm BP. However, patients with RSVA and a coexisting CoA present invariably with acute or subacute hemodynamic decompensation leading to a congested state. The rupture significantly increases the preload acutely while CoA results in chronic increase in afterload. Various strategies for management of these complex anatomical abnormalities include (i) single-stage surgical, (ii) two-stage surgical, and (iii) staged hybrid approach. Until recently, two-staged surgical approach is the most acceptable. This contributes to the improvement of the patient's clinical condition, with the further advantage of enabling safe perfusion during the second stage of the repair.

Fig. 2 – A. Aortic root angiography in left anterior oblique with cranial angulation showing RSVA arising from noncoronary sinus and draining into the right atrium. B. Noodle wire grabbed with a 15 mm Amplatz gooseneck snare to establish the arteriovenous loop. C. Lifetech Duct occluder (18 mm × 16 mm) with its attached cable positioned through the delivery sheath at the defect site D. Complete occlusion after duct occluder placement.
(RSVA closure), a few weeks later. As CoA is approached by lateral thoracotomy and RSVA by median sternotomy, a two-staged procedure is an acceptable option after a gap of 4–6 weeks.\(^9\) The consensus is that single surgical approach would increase excessively the risk of the procedure. A prerequisite for a single-stage repair is good biventricular function.\(^{10}\) In patients with left ventricle (LV) dysfunction, simultaneous repair is a double-edged sword. Earlier repair of RSVA can markedly increase afterload, and on the other hand, earlier repair of CoA can significantly decrease peripheral vascular resistance, decreased perfusion, and aggravation of heart failure. Hence, in cases with severe LV dysfunction, staged surgical repair or staged hybrid procedure or staged transcatheter approach could be a better option.

During transcatheter treatment, the sequence of intervention for the two lesions could be a matter of debate. Since RSVA is the “culprit lesion”, it would seem logical to address it first. However, since the aortic end of the RSVA was large (measuring 11 mm), a reason for one of the failures in our previous experience,\(^3\) we were not absolutely sure about the success of TCC. Indeed, after stenting of CoA, during the TCC of RSVA, we were unsuccessful in our first attempt with the 16/14 mm ADO. It has been our usual practice to use a device size 2–4 mm larger than the defect.\(^{11}\) Upsizing the device may be necessary at times because of the flimsy (“wind-sock”-like) margins of the defect. Hence we prioritized the stenting of CoA with the initial goal of at least avoiding two different surgeries through different incisions and minimizing the complexity of one-stage surgical procedure by dealing with one of them (CoA) by transcatheter approach. However, this approach of intervening on CoA first increased the “dwell-time” of a large bore 12F femoral arterial sheath with a potential for arterial complication. Indeed, there was increased bleeding, albeit mild, during catheter exchanges through the check valve of the long 12F sheath, and we had to exchange for another short 12F sheath, after accessing the ascending aorta through the venous side with the Amplatzer delivery sheath. The radial access was thus “well thought” not only for controlled angiography during TCC of RSVA, but also it could have proven valuable for establishing the arteriovenous loop without traversing the intervened stented CoA. Poststenting, there was an immediate significant fall in the ascending aortic pressures, which could have decreased the magnitude of shunt through RSVA. Patient remained stable and hemodynamic parameters were under control, so we went ahead with TCC of RSVA. To the best of our knowledge, simultaneous transcatheter intervention of both RSVA and CoA has not been attempted before in an acutely ill patient presenting with heart failure. In our largest reported series of 20 cases of TCC of RSVA, we have previously documented successful simultaneous transcatheter intervention for both these lesions in a young female, presenting with secondary hypertension due to CoA who was incidentally detected to have a small RSVA requiring the smallest ADO device (6/4 mm).\(^1\) In another case report,\(^{11}\) balloon dilatation of CoA and coil occlusion of a small RSVA has been staged six weeks apart in a 9-year-old boy, detected to have a cardiac murmur on routine examination.

4. Conclusion

In summary, there has been limited literature on transcatheter interventional treatment of RSVA with coexisting CoA. Although technically challenging, our experience suggests that TCC of RSVA and simultaneous stent implantation for CoA is a safe and effective alternative to a more complex surgery requiring two different approaches in hemodynamically unstable patient.

Conflict of interest

None.

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