

Percutaneous transcatheter closure of ruptured sinus of Valsalva aneurysm

Rüptüre sinüs Valsalva anevrizmasının perkütan transkateter yol ile kapatılması

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Summary– A sinus of Valsalva aneurysm (SVA) is a rare congenital anomaly. SVA rupture can lead to biventricular failure as a result of systemic to pulmonary shunting. Surgical repair has been the traditional treatment for these aneurysms. More recently, percutaneous treatment has been successfully performed on appropriate patients. This is the case of a 36-year-old man who had a ruptured SVA that was successfully treated using a catheter-based approach and a patent ductus arteriosus occluder device.

A sinus of Valsalva aneurysm (SVA) is a rare cardiac malformation most often caused by a congenital deficiency of muscular and elastic tissue in the aortic wall of the sinus of Valsalva. Periaortic inflammation, atherosclerosis, trauma, or aortic dissection can also cause this anomaly.^[1] As the rupture of an SVA is a life-threatening event, intervention is recommended as soon as the condition is diagnosed. Ruptured aneurysms in this location have been treated surgically for many years. Cullen et al.^[2] performed the first percutaneous closure in 1994, and since then percutaneous closure of an SVA has increasingly been used in appropriate cases.^[3] A successful transcatheter closure of a ruptured SVA is presented in this report.

CASE REPORT

A 36-year-old male patient was referred to a cardiology clinic because of a murmur heard in a physical examination. The patient had experienced fatigue and dyspnea for a year. A continuous grade 3/6 murmur was heard in the left third intercostal space on phys-

Özet– Sinüs Valsalva anevrizmaları ender doğumsal anomalilerdir. Valsalva sinüs anevrizması rüptürü nedeniyle sistemik akım ile pulmoner akım arasında oluşan şant, biventriküler yetersizliğe yol açar. Cerrahi onarım, bu anevrizmaların standart tedavidir. Son dönemde, uygun hastalara perkütan tedavi başarıyla uygulanmaktadır. Sinüs Valsalva anevrizma rüptürü olan ve patent duktus arteriosus okluder cihazı ile katetere dayalı bir yaklaşımla başarıyla tedavi edilen 36 yaşında bir erkek hastayı sunuyoruz.

ical examination. A transthoracic echocardiogram was performed, followed by a transesophageal echocardiogram (TEE). An SVA at the noncoronary sinus that protruded into the right atrium was observed. Color Doppler imaging revealed continuous flow from the aorta to the right atrium (Fig. 1a, b, Video 1*). A ruptured segment of the SVA 6 mm in diameter was noted. A pulmonary-to-systemic flow ratio (Qp/Qs) of 1.55:1 was calculated at the time of cardiac catheterization and a decision was made to pursue percutaneous closure.

Written consent was obtained from the patient and the corrective procedure was performed with fluoroscopy and TEE guidance and conscious sedation. The right femoral artery and vein were catheterized. The ruptured SVA and a pigtail catheter were visualized using aortography in the left anterior oblique 30° and cranial 15° view (Video 2*). A right Judkins

Abbreviations:

PDA	Patent ductus arteriosus
SVA	Sinus of Valsalva aneurysm
TEE	Transesophageal echocardiogram

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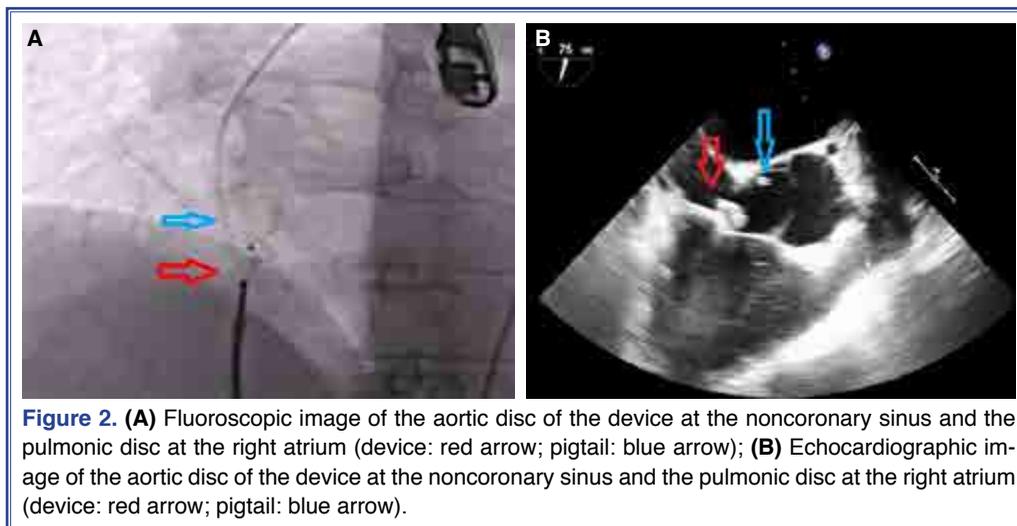
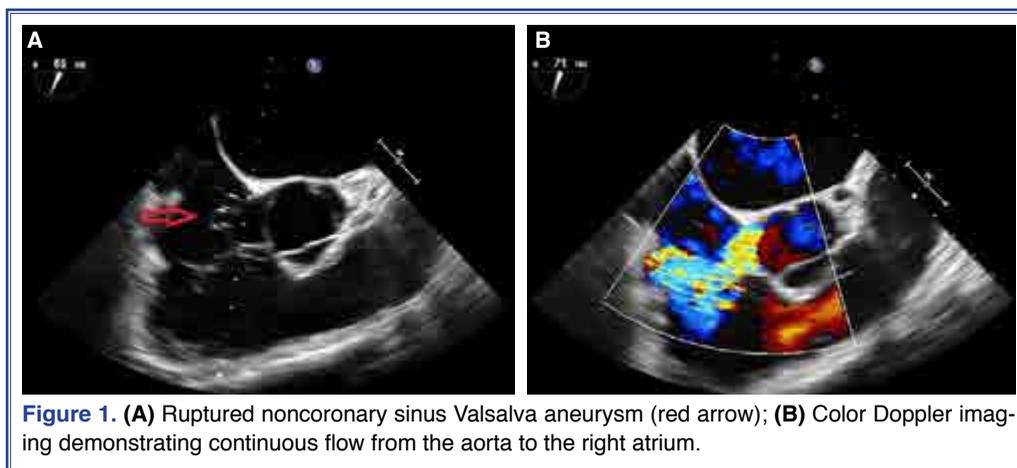
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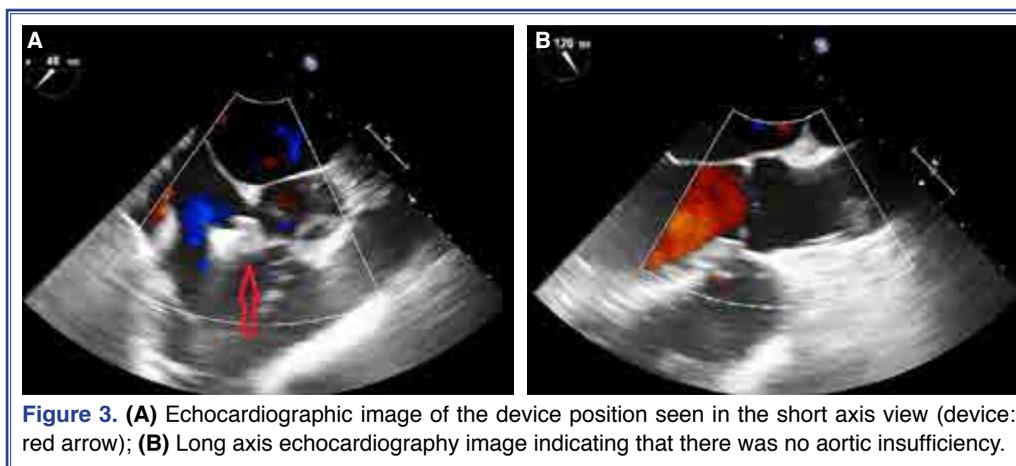
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coronary catheter was inserted through the right femoral artery and inserted at the noncoronary sinus. The ruptured SVA was crossed with the Judkins catheter. Next, a 300-cm exchange guidewire (Rope wire; AGA Medical Holdings Inc., Plymouth, MN, USA) was advanced through the ruptured SVA to the right atrium. It was caught with a snare (Goose Neck Snare; microvenes Main) and was removed through the right femoral vein. An arteriovenous loop was formed. A 7-F, 110-cm delivery system (Lepu Medical Technology (Beijing) Co., Ltd., Beijing, China) was advanced through the ruptured SVA to the ascending aorta over the wire from the right femoral vein. The exchange guidewire was removed after the appropriate position was reached in the ascending aorta (the tip of the delivery catheter was directed toward the ascending aorta). An 8/10-mm conical patent ductus arteriosus (PDA) occluder device

(Cone Shape PDA Occluder, Lepu Medical Technology (Beijing) Co., Beijing, China) was selected to close the ruptured SVA. The device was advanced to the end of the delivery catheter with a pusher catheter. The aortic disc of the device was deployed at the noncoronary sinus, followed by the pulmonary disc at the right atrium after confirming each step with fluoroscopy and TEE (Fig. 2a, b, Video 3^{*}). Once an optimal position in the ruptured SVA was confirmed, the device was deployed. There was no significant aortic regurgitation or any encroachment on coronary arteries seen on TEE or control aortography (Fig. 3a, b, Video 4, 5^{*}). The patient was discharged 48 hours after the procedure. Treatment for 6 months with acetylsalicylic acid 100 mg and clopidogrel 75 mg daily was initiated after the procedure. No residual defect was observed at the 6-month control examination.





DISCUSSION

An SVA is usually a congenital anomaly and accounts for 0.1% to 3.5% of congenital heart defects, with a greater incidence in Asians. An SVA is typically the result of a lack of fusion between the tunica media and the annulus fibrosus of the aortic valve. Aneurysms usually originate in the right coronary sinus or non-coronary sinus and terminate in the right ventricle and the right atrium. The aneurysm usually ruptures in the period between adolescence and early adulthood.^[4-6] The most commonly associated defects are a ventricular septal defect, bicuspid aortic valve, infundibular pulmonary stenosis, patent ductus arteriosus, atrial septal defect, coarctation and aortic regurgitation, or aortic stenosis.^[4]

The survival rate for congenital SVA patients is 95% at 20 years, since most SVAs do not rupture before 20 years of age. An unruptured SVA has been observed in serial monitoring up to several years after initial diagnosis, but most unruptured SVAs have been found to progress and ultimately rupture. A rupture can have serious hemodynamic effects, especially when it occurs quickly. About 35% to 58% of patients with a ruptured SVA become acutely symptomatic.^[6] The most common symptoms are dyspnea, orthopnea, palpitations, fatigability, palpitations, chest pain, and even sudden death.^[4] Adams et al.^[7] documented a mean survival period of 3.9 years in patients with an untreated ruptured SVA. Therefore, early closure is recommended as soon as the diagnosis is confirmed.

At least 2 orthogonal projections are needed for better visualization of the rupture site and the receiv-

ing chamber. Ruptures from the right or non-coronary sinus into the right atrium or with right ventricular inflow may be visualized in anteroposterior or right anterior oblique views (20–30°) with minimal cranial angulations. Ruptures from the right sinus into the right ventricular outflow tract or pulmonary artery are visualized in left anterior oblique views (30–40°) with cranial angulations of about 20–30°.^[8] TEE is also needed to determine the correct placement of the closure device and to manage possible complications during the operation.

Since 1956, the primary route of treatment has been surgical closure, but the associated morbidity is high as a result of the implementation of a sternotomy and cardiopulmonary bypass.^[6] A percutaneous closure technique is now available and has proven to be a simpler method with less morbidity. The first case of a transcatheter closure of a ruptured SVA was described in 1994 by Cullen et al.^[2] in a patient with recurrent sinus of Valsalva rupture after prior surgical repair. A Rashkind umbrella device was used to perform the closure.^[2] Fedson et al.^[9] reported the first case of the use of the Amplatzer ductal occluder device (ADO; St. Jude Medical, St. Paul, MN, USA) to repair a ruptured sinus of Valsalva aneurysm in 2003. Since then, the use of percutaneous closure has grown.^[4,10]

The most commonly used device is the ADO; other less frequently used devices included muscular ventricular septal defect occluders, atrial septal defect occluders, Rashkind umbrella devices, and rarely, coils.^[4] A venous antegrade closure technique is used for conventional ductal occluders and a retrograde aortic approach via the femoral artery is used for devices such

as muscular ventricular septal occluders and the ADO-II device (St. Jude Medical, St. Paul, MN, USA).^[8,11]

In a review published by Kuriakose et al.,^[4] the mean minimal diameter of the ruptured site was 7 mm (range: 2–15 mm), and accordingly, the device size ranged from 6/4 mm to 16/14 mm. The lesions were measured using TEE, and the authors selected devices that were 2 mm to 4 mm larger than the defect. In our case, the ruptured SVA diameter was 6 mm and we used an 8/10 mm MemoPart PDA occluder device (Lepu Medical Technology (Beijing) Co., Ltd., Beijing, China) for percutaneous closure. Devices should be released only after confirming precise placement, making certain there is no significant aortic regurgitation, tricuspid regurgitation, right ventricular outflow tract obstruction, arrhythmia, or coronary encroachment.^[4]

Potential complications of percutaneous closure of a ruptured SVA include valve regurgitation, coronary obstruction, right ventricular outflow obstruction, hemolysis associated with residual shunt, arrhythmia, and device embolization.^[3,4,8] Valve regurgitation, device embolization, right ventricular outflow obstruction, and coronary obstruction are usually associated with the need to use large devices to provide closure of a large ruptured SVA.

Approximately 3% of SVAs closed with a percutaneous technique have required a surgical procedure at the closure site.^[3,4,8] About 4% of SVAs treated surgically have required reoperation due to aortic insufficiency.^[3,4,8] In patients treated with surgery, mortality has been reported to be approximately 1.5% in the early period and 3% in the late period, but it is noteworthy that these data were obtained from studies conducted in 1960 to 2007.^[3,4,8]

In earlier SVA cases, postintervention anticoagulation typically included the administration of acetylsalicylic acid for 6 months, and sometimes clopidogrel for the first 4 to 6 weeks or for as long as 6 months.^[4,8]

Conclusion

The percutaneous closure of a ruptured SVA can be successfully performed in the appropriate patients. Percutaneous closure with the right device can be a reliable and safe alternative method to surgical treatment of a ruptured SVA.

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*Supplementary video file associated with this article can be found in the online version of the journal.

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Keywords: Patent ductus arteriosus occluder; ruptured sinus of Valsalva aneurysm; transcatheter closure.

Anahtar sözcükler: Patent ductus arteriosus tıkaçı; sinüs Valsalva anevrizma rüptürü; kateter aracılı kapama.