Ruptured Non-Coronary Sinus of Valsalva Aneurysm with Left Ventricular Outflow Tract-Left Atrial Fistula

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ABSTRACT

Aneurysm of the aortic sinus is a rare pathology and may remain clinically asymptomatic until rupture occurs. The rupture is seen most commonly into the right side of the heart but occasionally into the pericardium, the pulmonary artery, and the left ventricle. Only a few cases have been reported of rupture of a noncoronary sinus of valsalva aneurysm into the left atrium. We report a case of previously healthy 68 years old male who suddenly developed chest pain and pulmonary edema. Echocardiography and angiography revealed a rupture of a noncoronary sinus of valsalva aneurysm into the left atrium.
Key Words: Aneurysm of sinus valsalva; noncoronary sinus; left atrial fistula

Sol Ventrikül Çıkım Yolu ile Sol Atrium Arasında Fistül Oluşturan Rüptüre Non-Koroner Sinüs Valsalva Anevrizması

ÖZET

Aortik sinüs anevrizması nadir görülen bir patoloji olup rüptür oluşana kadar asemptomatik kalabilmektedir. Rüptür olursa sıklıkla kalbin sağ tarafına, zaman zaman da; perikardiuma, pulmoner artere ve sol ventriküle olabilir. Non-koroner sinüs valsalva anevrizmasının sol atriuma rüptüre olduğu ancak birkaç vaka rapor edilmiştir. Daha önce herhangi bir yakınması olmayan 68 yaşında erkek hastada ani gelişen göğüs ağrısı ve akciğer ödemi tablosu sonrası yapılan ekokardiyografi ve anjiografide non-koroner sinüs valsalva anevrizmasının sol atriuma rüptüre olduğu saptandı. Bu nadir durumu olgu sunumumuzda paylaşmak istedik.
Anahtar Kelimeler: Sinüs valsalva anevrizması; nonkoroner sinüs; sol atrial fistül

INTRODUCTION

Aneurysm of the aortic sinus is a rare disorder with an estimated prevalence of 1% in patients undergoing open-heart surgery[1]. Sinus of Valsalva (SV) aneurysms can be congenital or acquired. They may remain clinically silent until rupture causes symptoms related to compression of adjacent structures or intracardiac shunting[2]. If rupture occurs it is most commonly into the right side of the heart, but occasionally into the pericardium, the pulmonary artery, and the left ventricle. We report a case of previously healthy 68 years old male who suddenly developed severe chest pain and pulmonary edema. Echocardiography and angiography revealed an aneurysm of the noncoronary sinus of Valsalva which had ruptured into the left atrium.

CASE REPORT

A 68 year old male patient with no past medical history presents with a sudden onset dyspnea. He was referred to the Cardiology Department of our hospital. According to the transthoracic echocardiography (TTE) a ruptured non-coronary Sinus Valsalva Aneurysm (SVA) was showing passage to the left atrium and the left ventricle. Cardiac chambers were dilated (the left atrium diameter was 5 cm, the left ventricle diastolic diameter was 6.6 cm, the left ventricle systolic diameter was 4 cm). No Ventricular Septal Defect (VSD) was observed. The patient had mild aortic insufficiency, moderate tricuspid valve insufficiency and an ejection fraction of 60%. The pulmonary artery peak systolic pressure measurement was 55 mmHg. According to the coronary angiography, the coronary vessels were normal and injection into the aortic root lead to severe leakage into the left ventricle (Figure 1). The patient was referred to our clinic for operation. A standard bicaval cannulation procedure was performed and the left atrium was explored via a transseptal incision. The orifice of the aneurysm was seen on the non-coronary sinus of valsalva (Figure 2). The aneurysmal sac

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was ruptured. We found a defect (the aneurysmal sac was formed a fistula between left ventricle outflow tract and left atrium) at left ventricle outflow zone under the non-coronary cusp. The forceps inserted into the defect was passing into the left atrium (Figure 3). During the surgical procedure; we repaired the defect at left ventricle outflow by primary 4/0 sutures. Following aneurysmal sac resection, we closed the orifice of the aneurysmal sac using a pericardial patch. The intraoperative transesophageal echocardiography (TEE) revealed only a mild aortic insufficiency and a minimal passage from left ventricular outflow tract (LVOT)

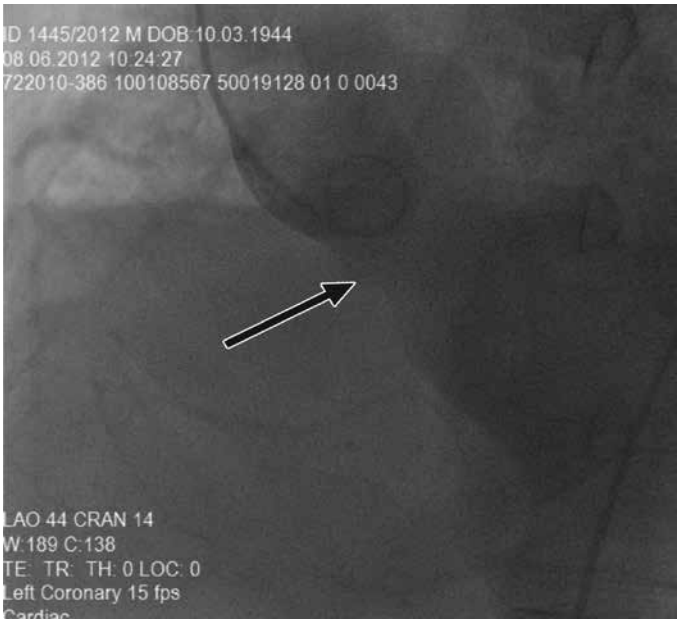

Figure 1. Preoperative aortic root injection (the arrow indicates aneurysm of the noncoronary sinus rupture into the left atrium)

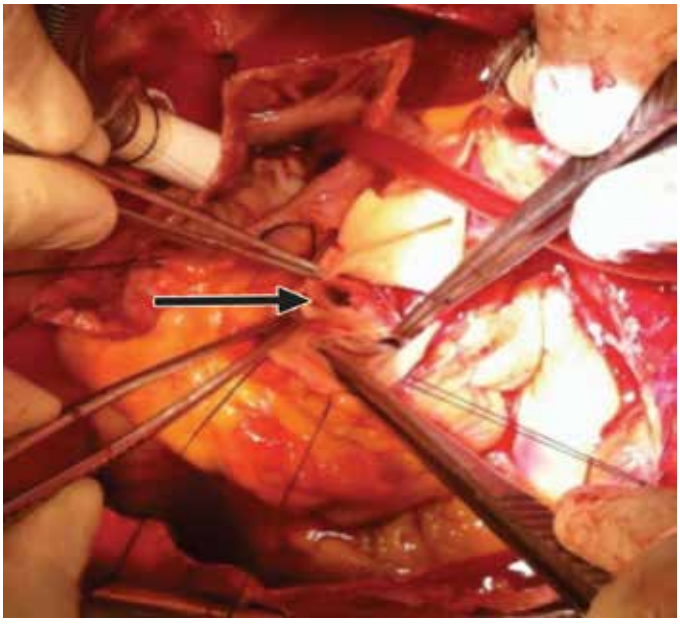

Figure 2. The arrow indicates orifice of non-coronary sinus valsalva aneurism sac

to the left atrium which was considered as the puncture points. The patient made an uneventful postoperative recovery, being discharged from hospital on the post-operative 9th day. The patient didn't have any clinical symptoms. Cardiac Magnetic Resonance Imaging (MRI) was performed 3 months later than the operation. The MRI images showed 2-3 mm connection from the LVOT to the left atrium (Figure 4). During the follow up period the patient had no complaints.

DISCUSSION

Aortic sinus aneurysms are rare disorders. They are most commonly congenital and although some cases associated with syphilis, infective endocarditis, trauma, atherosclerosis and aortic dissection have been described[3].

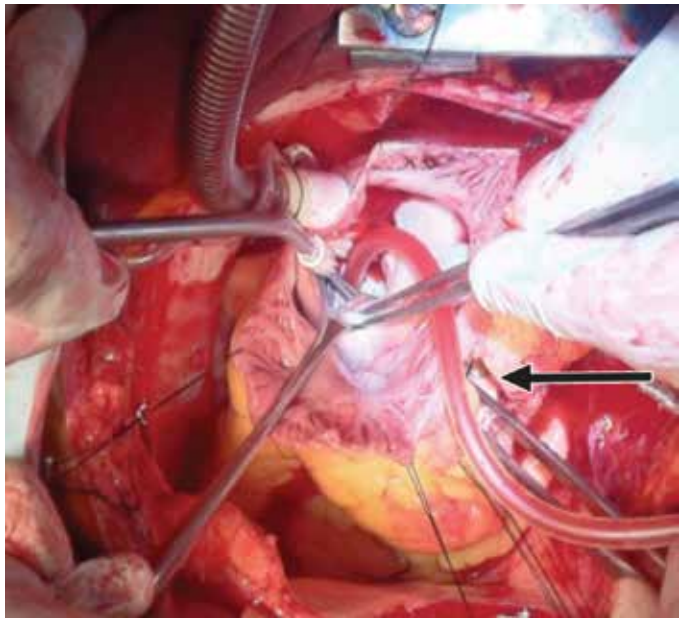

Figure 3. The arrow indicates the tissue forceps inserted into the defect was passed into the left atrium

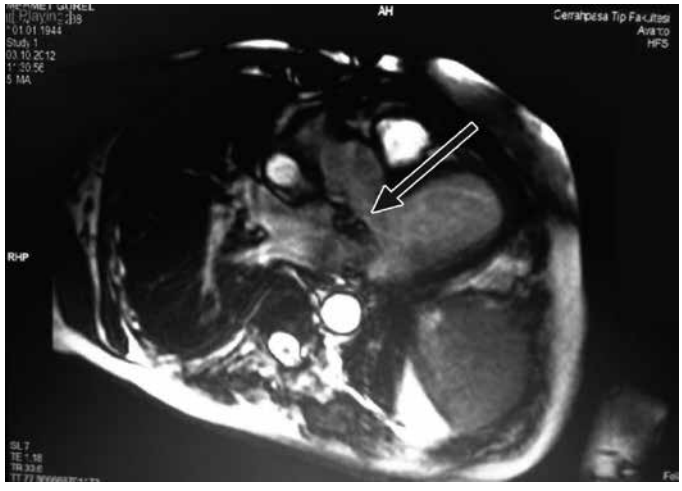

Figure 4. Two-three mm progression from the LVOT to the left atrium was observed from the MRI (black arrow)

The first formal classification system for sinus of Valsalva aneurysm was proposed by Sakakibara and Konno in 1962 according to the sinus affected and the location of their projection or rupture. A recent report suggested a modified Sakakibara classification.In the modified system, the etiology of the lesion is no longer emphasized; rather, the RSVA is classified by the anatomic location of the protrusion site. According to this modified system our patient is with lesion 5 (type 5: Other rare conditions eg, rupture into left atrium, pulmonary artery, left ventricle, or other structures)[4].

The aneurysms usually arise from the right coronary sinus (65-85%), less frequently from the noncoronary sinus (10-20%) and rarely from the left coronary sinus (<5%).

If they do not rupture, the aneurysms can present as merely echocardiographic findings or, less often, they are revealed through compression of neighbouring structures, leading to impaired coronary circulation, aortic or tricuspid valve dysfunction, or right ventricular outflow tract obstruction[5].

Clinical presentation depends on the time since rupture, the chamber into which the fistula opens, the flow volume and complications such as aortic or mitral valve regurgitation, atrioventricular block, endocarditis or cardiac tamponade. Rupture of a non-coronary sinus aneurysm typically results in the formation of a fistulous tract with the right atrium (RA), while rupture of right coronary sinus aneurysm frequently leads to fistulous communication with the right ventricle[6]. On the other hand, rupture within the left atrium may be seen in the left sinus valsalva aneurism which is a very rare condition[7].

As far as we know, only few cases have been reported of noncoronary of sinus valsalva rupture into the left atrium[8]. In our case, echocardiography and angiography revealed an aneurysm of the noncoronary sinus of Valsalva which had ruptured into the left atrium. This was confirmed at the operation and it was noted that there were no signs of bacterial infection or rheumatic valve disease. We, therefore, concluded that the aneurysm was of a congenital origin.

In case of rupture, the most common manifestation is heart failure, varying greatly in intensity and speed of progression. Rupture of SV aneurysms is associated with high mortality, with survival of less than four years and surgical repair is therefore indicated. More recently, some alternative treatment techniques have been reported. Successful percutaneous treatment of ruptured SVA using a duct occlusion device and with coil embolization have been described[9,10].

The resection of the aneurysmal sac and closure of the defect is possible transaortically especially for the unruptured ones. But we want to emphasise that for the ruptured aneurysms a combined approach through aortotomy and the involved chamber will be more helpful. Either direct or patch closure can be used to close the rupture hole. If the diameter of the ruptured SVA fistula is less than 1.0 cm and the orifice edge is solid it can be closed directly without a patch. Resection of the aneurysm sac should be small and intraoperative stretching must be gentle to avoid affecting the aortic ring or damaging the valve[11]. In our clinic this case was the first recorded SVA case ruptured to the left atrium, however repairing the defect at left ventricle outflow by using a patch or pledgeted sutures instead of primary 4/0 sutures would have better outcomes for preventing passage from left ventricular outflow tract (LVOT) to the left atrium which was considered as the puncture points. Once the diagnosis of ruptured SVA is confirmed, surgical correction should be performed immediately and if not an emergent operation, necessary operation should not be delayed because of concerns about cardiac function improvement. The strategy of operation should be based on the location, size of the fistula, presence or absence of VSD and aortic insuffiency, and other possible associations[11].

In conclusion, surgery is the definitive therapy and urgent surgical repair is recommended in all patients with ruptured SVA, especially with intracardiac shunting.

CONFLICT of INTEREST

The authors reported no conflict of interest related to this article.

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