

CASE REPORTS

Ruptured Sinus of Valsalva Aneurysm: A Rare Cause of Sudden, Unexpected Death

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ABSTRACT

Introduction

A sinus of Valsalva aneurysm (SOVA) is a rare dilatation of the aortic wall. Predominantly a congenital lesion, it results from a failure of tissue planes to properly fuse to form the sinus of Valsalva and semilunar cusps of the aortic valve. SOVAs are typically discovered incidentally and most commonly involve the right sinus. Rupture of this aneurysm can cause sudden death via cardiac tamponade or hemodynamic derangements causing cardiovascular insufficiency. This report describes two cases of ruptured SOVAs that resulted in sudden, witnessed collapse and death.

Case Presentations

Case One

A man in his early 30s suddenly collapsed while walking with his girlfriend, after telling her that he did not feel well. He was transported to a local medical center but resuscitative efforts were unsuccessful. He had no personal or family history of heart disease, stroke, aneurysms or sudden, unexplained death at an early age. Because the death was sudden and unexpected in someone of relatively young age, the death fell was investigated by the regional Medical Examiner's Office, associated with the Department of Pathology at Wake Forest School of Medicine. At autopsy, a ruptured thin-walled aneurysm 4 cm in size was seen arising from the right sinus of Valsalva and protruding into the aortic outflow tract (Figure 1). The aneurysm was connected

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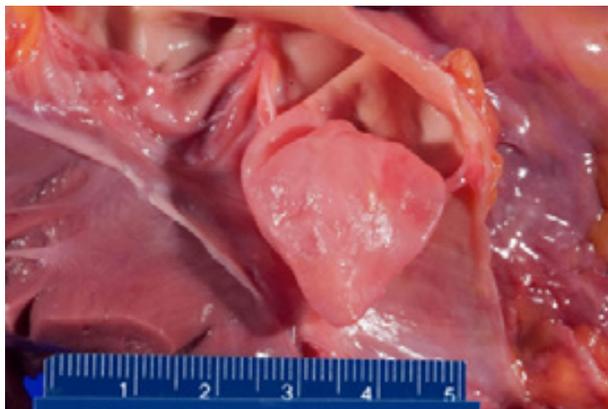


Figure 1. The aneurysm forms a tongue-like protrusion, hanging over the aortic cusp.

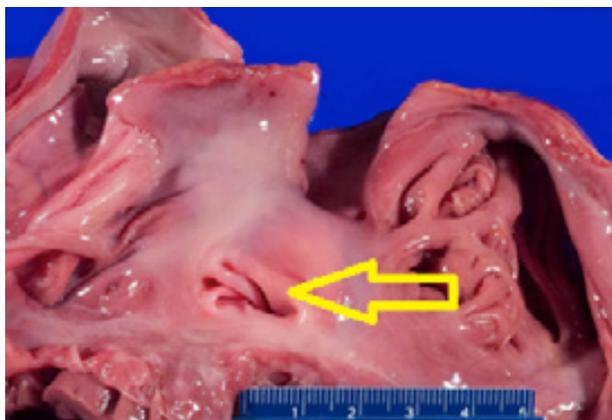


Figure 2. The opening of the fistula (arrow) seen within the right atrial wall.

to the right atrium by a fistula (Figure 2). The rupture created hemodynamic instability with the formation of a left-to-right shunt from the aorta to the right atrium of the heart. The cause of death was cardiovascular insufficiency as a result of the shunt created by the ruptured SOVA aneurysm.

Case Two

A young man in his early 30s had a witnessed sudden collapse while doing non-strenuous labor at his place of employment. Bystanders initiated cardiopulmonary resuscitation until emergency medical technicians arrived, but he was pronounced dead at the scene. He had no significant personal or family history of cardiac disease or risk factors for death at an early age. Since the death was sudden, unexplained and occurred at the workplace, this case also was investigated by the Medical Examiner's Office. At autopsy, the decedent was obese and had no external evidence of trauma. A tense, distended pericardial sac was filled with 150 ml of blood, which would have produced cardiac tamponade. The source of the blood was an aneurysm arising from the left sinus of Valsalva that had bulged outward toward the epicardial surface of the left atrium, directly underneath the path of the left main coronary artery. The left coronary artery was displaced along the contour of the aneurysm, with a perforation measuring less than 0.1 cm in the wall of the aneurysm (Figure 3).

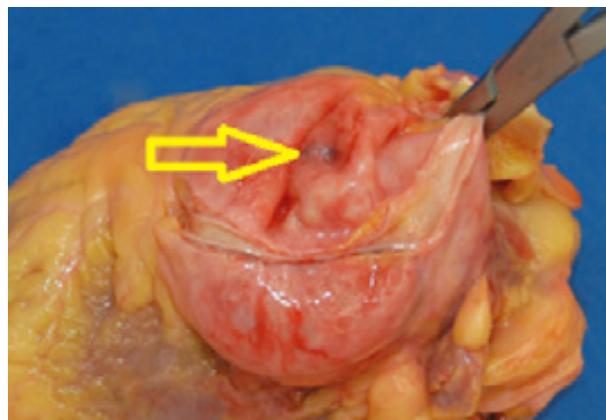


Figure 3. The left main coronary artery running horizontally over the aneurysm, and the perforation (arrow) seen on the surface of the aneurysm.

Discussion

A sinus of Valsalva aneurysm (SOVA) is a rare cardiac lesion. A study of 270 archived hearts over a 13-year period uncovered just one ruptured sinus of Valsalva aneurysm.¹ These aneurysms can be congenital or acquired, although the congenital type is more common. The congenital type of SOVA is likely due to weakness at the junction of the aortic media and the annulus fibrosus, and they are most commonly associated with congenital ventricular septal defects.² The acquired type of SOVAs are most likely to be the result of conditions weakening the aortic wall: syphilis, endocarditis, atherosclerosis, or connective tissue disorders.² Relatively high aortic pressures contribute to the dilation of the aneurysm, until it either ruptures or creates an obstruction or compression of adjacent structures.³ The SOVA can occur in any of the three sinuses, but usually occurs in the right sinus. It is rarest in the left sinus; Smer et al documented only 30 such cases as of 2015.³ The onset of symptoms most likely occurs between 20 to 30 years of age.⁴

Unruptured SOVAs can be asymptomatic or may lead to blockage of coronary blood flow and ischemia. Displacement of the left main coronary artery can occur from an unruptured SOVA and can result in cardiogenic shock.^{5,6} One case of a non-coronary SOVA that ruptured into the inter-atrial

septum has been described in the literature.⁷ Rupture may be spontaneous or incited by factors such as increased pressure within the proximal aorta (i.e., in increased diastolic pressure) or chest wall pressure (i.e., trauma). Zhu et al described a ruptured sinus of Valsalva aneurysm caused by blunt force trauma to the chest.⁸ These aneurysms, once ruptured, can create a continuous, mechanical-sounding murmur from a distinctly lower position than the similar-sounding patent ductus arteriosus type of murmur.⁹ A ruptured SOVA can result in cardiac tamponade, or form a fistulous connection with one of the cardiac chambers; this situation requires immediate surgical intervention.⁹

When the aneurysm produces cardiac symptoms rather than sudden death, diagnosis is classically made with either transthoracic or transesophageal echocardiography. However, recent advances in CT technology have allowed better imaging with motion-free images and faster acquisition time.¹⁰ Standard treatment has been surgical repair via a cardiopulmonary bypass, but recently, a report of a transcatheter closure procedure has shown definitive advantages, by minimizing risk.¹¹ There are no screening tools for pre-symptomatic SOVAs, and they continue to be more commonly identified incidentally.

Conclusion

A sinus of Valsalva aneurysm, which is often undiagnosed during life, is a rare cause of sudden death. If unruptured, these aneurysms (depending on their location) can impede aortic blood flow, displace coronary arteries, or remain asymptomatic. Once ruptured, these aneurysms can result in immediate hemorrhage, causing cardiac tamponade or cardiovascular insufficiency, again depending on the location.

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