

Case Report

Rupture of a Non Coronary Sinus of Valsalva Aneurysm in Side the Right Atrium

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INTRODUCTION

Abnormalities of the structures of the aortic ring and especially of sinus of Valsalva are rare and when present are usually congenital. Aneurysms of sinus of Valsalva (SVAs) consist of 0,5 -3% of all congenital heart defects(1) and are present at 0,15-1,5% of all patients who undergo open heart surgery(2). Acquired aneurysms are even rarer, but they are more invasive and occupy multiple sinuses. Most commonly occupy the right or non coronary sinus(3): approximately 65-85% of SVAs originate from the right sinus of Valsalva, 10-30% originate from noncoronary sinus while aneurysms originating from the left sinus are extremely rare (<5%). (4) Symptoms may appear in young people (even in childhood) free of medical history, with no cardiovascular risk factors (5), while most cases which concern ruptured aneurysms occur from puberty until age of 30 years and are often diagnosed or present clinical symptoms around this age (4). In any case, presence of SVAs from infancy to seventh decade of life has been described(2). Their incidence in males is 2-3 times higher than in females. Acute increase in the size of the aneurysm may be life threatening, while its rupture is usually a fatal event and commonly occurs in the right chambers of the heart (2).

The archives of the Laboratory of Forensic Sciences at the University of Crete have been reviewed for the last decade (2001-2011) and only one case of natural death due to rupture of sinus of Valsalva's aneurysm has been recorded.

CASE REPORT

A 53-year old man collapsed during a family reunion, after complaining for an acute chest pain. He was transported at the emergency room, where he was pronounced dead. Information provided by his next of kin revealed no previous medical history. The body was transferred to the morgue for routine postmortem examination.

Autopsy findings: After his clothes were carefully examined, resulted that the deceased carried no medication in his pockets. The external examination of the body showed no evidence of any type of injury. The ankles appeared edematous, while toes were covered by dermal ulcers. In the body cavities, no fluids or haemorrhage was observed. The heart weighted 560 grams and appeared enlarged, while the right chambers seemed dilated. Opening the right atrium, we located a saccoid aneurysm of non coronary sinus of Valsalva (4X5 cm) (Fig 1).

A linear rupture of 2,7 cm length was also observed

on the upper surface of the aneurysm (Fig 2). The walls of both right and left ventricle, as well as the intraventricular septum were fattened, while coronary arteries were free of any pathological changes. The liver appeared cirrotic, while the macroscopic examination of organs revealed congestion.

Histological samples were taken for the routine microscopic examination. Cross sections of the aneurysm confirmed its atherosclerotic origin and the existence of necrotic material, haemorrhagically infiltrated, indicating a recent rupture.

Toxicological results didn't reveal alcohol consumption nor a drug induced death. The death after careful coestimation of all findings during autopsy, histological and toxicological analysis was attributed to the automatic rupture of a saccoid



Fig 1 The aneurysm is extended in the right atrium.



Fig 2 Appearance of the ruptured aneurysm while opening heart cavities



Fig 3 Necrotic material after dissecting the aneurysm

aneurysm of non coronary sinus of Valsalva and the manner of death was natural.

DISCUSSION

Aneurysms of Valsalva sinus are an acquired abnormality of the aortic ring. The congenital form of the disease is based on the lack of continuity or other deficiencies concerning the structure of the the aortic wall.. Abbott and Edwards and Burchell also have suggested that these lesions are formatted due to a structural deficiency in the media , related to the development of the distal bulbar septum(6). Otherwise its formation, may be due to trauma(7), inflammatory diseases, such as syphilis and tuberculosis(8) and connective tissue disorders (3) endocarditis (9), Marfan' s syndrome, Behcet's or degenerative diseases (10) or senile type dilatation of the aortic root (6). Atheromatosis of the aortic media may also contribute to aneurysm formation, as in the case presented (11)

Also, SVAs may be associated with congenital cardiac anomalies, including pulmonary stenosis ,atrial septal defect, bicuspid aortic valve, tetralogy of Fallot or coarctation of aorta (12). The aneurysms are usually located in the right chambers of the heart, however when occur in the left chambers, an

Table 1: Classification for SVA proposed by Sakakibara and Konno

Type I	Connect the right SV and the existing tract of the RV below the pulmonary valve
Type II	Connect the right SV and the VD in the supraventricularis crest
Type IIIa	Connect the right SV and the RA
Type IIIv	Connect the posterior zone of the right SV and the RV
Type IIIa+v	Connect the right SV and both RA and RV
Type IV	Connect the noncoronary SV and the RA

extracardial mass is formed (6).

According to their size and location aneurysms are divided into subtypes I-IV, according to the classification proposed by Sakakibara and Konno, since 1962 (Table 1) (13).

Although, a new and simplified classification was recently proposed by Guo HW et al, for non-coronary sinus of Valsalva aneurysm, dividing them into two types, type I: rupture or protrusion into right atrium; and type II: rupture or protrusion into right atrium or right ventricle near or at the tricuspid annulus (14). Symptoms arising due to aneurysms presence vary and may be associated with right heart failure (cyanosis and partial respiratory insufficiency) while when rupture occurs may range from asymptomatic murmur or end up in cardiogenic shock. It has been well described an interesting case of AV block III, attributed by the authors either to the external compression of coronary arteries or to wall distension from the aneurysm in the area of the right ventricle (1). Alternatively, an episode of rupture may be followed by intracardial shunting, when a communication is established with the right

atrium (Gerbode defect) or directly into the right ventricle (15,16). During the past decade, there were eight published cases in which a nonruptured aneurysm manifested with myocardial ischemia or infarct resulting from compression or occlusion of a coronary artery. (17-19)

Diagnosis is often set after random control, where a mass is revealed in a transthoracic echographic examination, while another advanced, reliable and accurate examination is the magnetic resonance imaging with the phase contrast method (20). The advantages of performing MR imaging in the setting of a known or suspected Valsalva sinus aneurysm include the ability to evaluate the left ventricular hemodynamic pattern, identify aortic regurgitation, and quantify any aortocardiac shunt or turbulent or fistulous blood flow. (19,21) Although angiography is considered the reference standard for confirming the presence of a Valsalva sinus aneurysm, most are initially seen at color Doppler echocardiography. Although in most cases CT was performed to confirm echocardiographic findings, the use of CT to help diagnose Valsalva sinus aneurysms may become more common as it becomes increasingly popular in urgent care settings. The high contrast resolution of CT also may make it possible to delineate an aortocardiac shunt, if present, and to identify a ruptured aneurysm by depicting a jet of contrast material extending from the aneurysm into the adjacent cardiac chamber (22). ECG-gated contrast material-enhanced multisection CT provides much better spatial resolution of cardiac structures than that attainable with other imaging methods. It also provides detailed anatomic depiction of Valsalva sinus aneurysms and surrounding cardiac structures (23)

The treatment of choice in such cases is excision of the aneurysm, a low risk operation with excellent

results in current cardiovascular surgery trends. Indications in order to perform a surgery include malignant arrhythmias, infection, acute ostial coronary artery obstruction and right ventricular outflow tract obstruction (2,24). An unofficial indication for surgery excision is also the enlargement of the aneurysm, when exceeds 50% of normal Valsalva sinus size. When atherosclerotic damages coexist in coronary arteries, it is imposed that any surgical treatment includes them too, because is correlated with high mortality rates (25). Additionally, a therapeutic approach is proposed by Guo HW et al, according to aneurysm's type. For type I, the right atrium approach is chosen, using direct suture with patch repair. For type II, the transaortic approach with right atrium incision is chosen, with patch repair through an aortic incision and direct suture through a right atrium incision.(14)

Recent interventional treatments in cases of ruptured aneurysms propose the occlusion of the defect with ASD closure devices or catheter closure.(4,5). Aneurysms which have undergone rupture should be treated with patch rather than a simple closure, especially the large ones. More radical treatment is the replacement of the root of the aorta and the ascending aorta as well, following reimplantation of coronary arteries, especially in cases where coexists dilated annulus or multiple sinus are involved (6).

Considering the frequency of ruptured aneurysms, a Web-based search of all clinical and surgical cases of Valsalva sinus aneurysms published in the English language over the past 10 years yielded a total of 1121 patients studied in vivo. 66% of them presented with a ruptured aneurysm and 34% not.(26)

Once rupture has occurred and not repaired, the mean survival is 1-4 years approximately (6). The mortality during early postoperative period is very low. The postoperative survival mentioned in Wng Z.J et al

s project is referred as a 90% for 10 years and 93% for a 20 year survival. (25), while in Takach T.J et al's study, the postoperative survival from a ruptured aneurysm is referred as high as 63% for a 10 year survival, while in unoperated cases the survival is 3,9 years after rupture. (6) One of most dangerous for patient's life complication is the automatic bleeding from anticoagulative treatment (20).

CONCLUSION

The aortic aneurysm is a rather common cause of sudden natural death, especially in the third age, and not rarely is due to atherosclerotic complications. The rupture is usually spontaneous inside the pericardiac sac. This ectopic growth of the aortic aneurysm inside the right atrium is an extremely rare phenomenon and consists an unusual heart abnormality. If they remain undiagnosed, they may lead to sudden death due to a sudden cardiac blockage from the rupture of their wall inside the cavity of the atrium and the short circuit of blood circulation in the ascending portion of the aorta circulating the blood to the right atrium back rather than the thoracic part and the rest arterial circulation. Sudden onset of a cardiac murmur should always include the differential diagnosis of a ruptured aneurysm of Valsalva sinus.

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