A Literature Review on Aneurysms of the Sinus of Valsalva

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Abstract

Sinus of valsalva aneurysm (SOVA) is a rare cardiac anomaly that is most commonly seen in the Asian population. It generally affects the right coronary sinus, with the most common complication is rupture into any cardiac chamber. Early surgical or percutaneous repair should be considered to prevent the condition from worsening and the development of complications.

Keywords: Aneurysm; Repair; Rupture; Sinus of Valsalva; Transthoracic Echocardiography; Transesophageal Echocardiography

Introduction

Sinus of valsalva aneurysm (SOVA) is an abnormal dilatation of the aortic wall placed between aortic valve and sinotubular junction resulting from a lack of continuity between the aortic media and aortic valve [1]. It is a rare cardiac anomaly occurring at an incidence of < 0.1% that is most commonly seen in the Asian population [2]. Most cases are congenital, but the disease can also be acquired in cases of trauma, infection, or degenerative disorders [3]. It generally affects the right coronary sinus (94% of the cases) and is less frequent in the noncoronary sinus (5%). it rarely affects the left coronary sinus (1%) [4,5]. The most common complication is rupture into the right atrium (29%) or ventricle (60%), with rupture into the left chambers (the left atrium [6%], left ventricle [4%]) or pericardium (1%) occurring rarely [4]. Rupture into PA or interventricular septum is very rare [5,6].

Aim of the Study

The aim of current study was review of the literature on the occurrence, clinical presentation, diagnosis and management of patients with ruptured sinus of valsalva aneurysm.

Evidence Acquisition

We did a literature search by PubMed and Google Scholar databases. PubMed MeSH terms encompassed issues including Sinus of Valsalva/abnormalities, Sinus of Valsalva/pathology, Sinus of Valsalva/therapy, Sinus of Valsalva/surgery and Sinus of Valsalva/injuries. Main keywords included sinus of Valsalva; aneurysm; repair and rupture.

Pathophysiology

A SOVA is a thin-walled outpunching of the aortic root, usually of a single sinus, between the aortic valve annulus and sinotubular ridge [7,8]. It can be caused by congenital or acquired abnormalities by an incidence of 0.09% of the general population, men are more affected (4:1) and is 5 times more common in Asian patients than in Caucasian. Acquired forms of SVA are caused by syphilis, infective endocarditis, atherosclerosis, cystic medial necrosis and trauma [9]. The rupture results in a fistulous communication of the involved aortic sinus into any cardiac chamber [8,10]. The most commonly involved sinus is the right coronary sinus, followed by the noncoronary sinus, and least common in the left sinus [8]. The right coronary sinus is adjacent to the RV outflow tract, membranous portion of the interventricular septum and anterior of the right atrium [11]. Aneurysms of the right sinus usually ruptured into right heart chambers [12]. The non-coronary sinus is near the base of the interatrial septum and lies next to both atria, the atrioventricular node and His bundle. The base of the left coronary sinus is adherent to the anterior wall of the left atrium and is covered by the pericardium [11]. Rupture usually occurs in the right ventricle, followed by the right atrium, with the least common location in the left ventricle, pulmonary artery, pericardium or interventricular septum [4,9]. The most common coexisting cardiac anomaly is ventricular septal defect occurring in 30-50% of patients with ruptured aneurysm. The second most common associated lesion is aortic regurgitation presenting in 20 - 30% of patients. Other uncommon coexisting cardiac anomalies including atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, aortic coarctation and subaortic stenosis [9].

History

Hope first reported congenital ruptured sinus of Valsalva aneurysm in 1839, and its clinical features was first described by Thurman in 1840 and echocardiographic findings was first reported by Rothbaum, et al. in 1974 [11]. Lillehei reported the first successful surgical treatment by cardiopulmonary bypass and interrupted silk stitches from involved cardiac chamber in 1957 and percutaneous closure of ruptured sinus was first attempted by Cullen., et al. in 1994 by Rashkind umbrella device in a patient who had previous surgical repair [13,14].

Clinical presentation

Ruptures usually occur between 20 - 40 years of age. The size, location and speed at which ruptures occur are major determinants of clinical outcome. The most common symptoms include chest pain, abdominal pain, and dyspnea. Patients may also have symptoms of acute heart failure, cardiac tamponade, hemodynamic instability and sudden cardiac death, with rupture into the interventricular septum causing left ventricular outflow tract obstruction. Non-ruptured SOVAs are usually asymptomatic. However, some patients may experience dyspnea, palpitations, angina pain, arrhythmia, atrial fibrillation, or complete heart block. Large SOVAs can be a nidus for thrombus formation leading to ischemic cardiac disease by obstructing epicardial coronary arteries. Aortic regurgitation occurs in 30 - 50% of patients who need an aortic valve replacement in addition to operative repair of ruptured or un-ruptured aneurysm. On physical exam, a loud superficial continuous murmur can be heard in patients with large or ruptured aneurysms [8].

Diagnostic imaging

Transthoracic echocardiography is the first-line imaging modality for the detection of SOVA, with transesophageal echocardiography being the diagnostic tool of choice. Echocardiographic study with color Doppler will appear continuous flow in systole and diastole, because the high pressure aorta is draining into a low pressure chamber (Figure 1-6). Magnetic resonance imaging is a useful complementary test for the evaluation of intracardiac shunts in ruptured SOVAs and Cardiac CT imaging has allowed to determine even small SOVAs. These two modalities have been used as supplemental or confirmatory tests [8].

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Figure 2: Transesophageal two-dimensional echocardiography and color Doppler flow velocity mapping in the midesophageal long-axis view (124 degree) revealed continuous turbulent flow (arrow) from the right coronary sinus aneurysm toward the RA (LV: Left Ventricle; LA: Left Atrium; RVOT: Right Ventricular Outflow Tract; AO: Aorta).

Figure 3: Transesophageal two-dimensional echocardiography in the mid-esophageal view (44 degree) demonstrated a ruptured left coronary sinus aneurysm (arrow) protruding into the main PA (pulmonary artery).

**Figure 4:** Transesophageal two-dimensional echocardiography and color Doppler flow velocity mapping in the midesophageal view (44 degree) revealed continuous turbulent flow from the left coronary sinus aneurysm toward the main PA.

**Figure 5:** Transesophageal two-dimensional echocardiography in the mid-esophageal long-axis view (126 degree) showed a ruptured noncoronary sinus aneurysm (arrow) into the LA (LV: Left Ventricle; LA: Left Atrium; RVOT: Right Ventricular Outflow Tract; AO: Aorta).

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Management

The mean survival in patients with an untreated ruptured SOVA is 1 - 2 years, indicating the need for early surgical intervention [14]. A non-ruptured SOVA may cause outflow tract obstruction, arrhythmia, or infection that an intervention is needed. Furthermore, surgical intervention is recommended in patients with an aneurysm diameter greater than 5.5 cm. Surgery should also be elected in those with bicuspid aortic valve and an aneurysm diameter greater than 5 cm or patients with connective tissue disorders and aneurysm > 4.5 cm, or a growth rate of > 0.5 cm/year [15]. Different surgical approaches have been introduced, with cardiopulmonary bypass and cardioplegic arrest needed in all cases. Three common approaches exist: 1) via the aortotomy and aortic root, 2) via the cardiac chamber in which the aneurysm has ruptured, or 3) a combined approach using both an aortotomy and an incision into the involved cardiac chamber. Two main closure techniques exist including primary closure and patch closure with former has been used for the repair of small SOVAs and latter for repair of larger ones because primary closure in those with larger aneurysms may involve the aortic sinus and culminate in the aortic valve incompetence, or may cause enormous tension of the tissue in the site of surgical repair resulting in a late recurrence of rupture. Overall operative mortality is 1-9-3.6% and 15-year survival rates are 90%. Early surgical repair should be considered to prevent the condition from worsening and the development of complications [8]. On the other hand, many recent reports have demonstrated favorable clinical outcomes by transcatheter closure devices as the surgical alternatives without any serious complications including embolization of the device, residual shunt, right ventricular outflow tract obstruction, worsening aortic regurgitation, or aortic tear [16].

Conclusion

SOVA is a rare cardiac anomaly which may result in rupture of aneurysm into any cardiac chamber. Early surgical or percutaneous repair should be considered to prevent the condition from worsening and the development of complications.

**Figure 6**: Transesophageal two-dimensional echocardiography and color Doppler flow velocity mapping in the midesophageal long-axis view (126 degree) demonstrated continuous turbulent flow (arrow) from the noncoronary sinus aneurysm toward the LA (LV: Left Ventricle; LA: Left Atrium; RVOT: Right Ventricular Outflow Tract; AO: Aorta).

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Bibliography


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