Rupture of Sinus of Valsalva Aneurysm (Type V Sakakibara Type) Treated by the Amplatzer PDA Occluder Device

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Abstract

Rupture of sinus of valsealva aneurysm is a rare anomaly. It is classified based on the site of rupture. The rarest type is an aneurysm rupturing into the left atrium, left ventricle, or pulmonary artery. The surgery is considered to be the treatment of choice in most of the cases where as trans-catheter treatment by the vascular occluder devices is an effective alternative method of treatment. We report the rarest variety of Sakakibara type V aneurysm treated by percutaneous transcatheter Amplatzer PDA occluder device with no post-procedural complication.

Introduction

Sinus of valsealva aneurysm (SVA) is defined as the enlargement of one or more of the sinuses between the aortic valve annulus and the sinotubular junction [1]. Focal expansions which form the walls of the aortic root are called the sinuses of Valsalva from which the left and the right coronary arteries arise from the respective coronary sinuses [2,3]. The non-coronary sinus does not give rise to any coronary artery. Thurman, in 1840, described the sinus of Valsalva aneurysm [4]. Aneurysms of the sinus of Valsalva are very uncommon, with an incidence ranging from 0.1 to 3.5% of all congenital heart defects. Men are affected more than women with a male–female ratio of 4:1 [5]. Aneurysms commonly originate from the right coronary sinus (70–90%), the noncoronary sinus (10–20%), and rarely from the left sinus (<5%) [1,5]. SVAs are usually clinically silent when there is no rupture, but the greater availability of diagnostic imaging has led to an increased incidental discovery of the lesion on echocardiography or cross-sectional imaging performed [5].

Case Report

Thirty three years old male patient, a known case of varicose veins of the lower limbs on follow up, presented with breathlessness and fatigueability and was misinterpreted as dilated cardiomyopathy with left ventricular dysfunction elsewhere and the patient was referred to our hospital for further work up. On clinical examination, the patient had a long end diastolic and short end systolic murmur at the base. Hence, the patient was subjected to a pre-operative echocardiography which revealed a sinus of Valsalva aneurysm with left ventricular ejection fraction of 25%. After the incidental detection, CT coronary angiography was performed for the confirmation of the anomaly, exact delineation and the subtyping.

Coronary CT angiography was performed on 128-row multi-detector CT scanner (Ingenuity, Philips); 80 ml of 350 mg I/ml non-ionic contrast injected through a dual-head Medrad pressure injector at a flow rate of 5.5 ml/s, using 100 kV, 570 mAs, 0.8-mm slice thickness, pitch 0.2, rotation time 0.4ms with ECG synchronization and retrospective gating.

CT coronary angiography axial, coronal and sagittal images showed a contrast filled out-pouching arising from the left coronary sinus suggestive of aneurysm. The wall of which showed calcification suggestive of atherosclerotic changes and the chronicity (Figure 1A) and the origin of the aneurysm and its relationship with the coronary arteries was clearly delineated (Figure 1B, 1D). The aneurysm showed communication with the left ventricle (Figure 1C, 1D), left atrium (Figure 1E) and the leak of contrast suggestive of rupture (Figure 1F) and was classified as type V Sakakibara ruptured SVA. Volume rendered images depicted the sinus of valsealva aneurysm arising from the left coronary sinus, origin of the left anterior descending artery, stretched out left circumflex artery and the communication between the aneurysm and the left atrium.

Figure 1: CT coronary angiography axial, coronal and sagittal images showing the aneurysm from the left coronary sinus which shows calcification in the wall of the aneurysm (A) and the origin of the aneurysm (B,D), its communication with the left ventricle (C, D), left atrium (F) and the leak of contrast suggestive of rupture (F).

Figure 2: CT coronary angiography volume rendered images showing sinus of valsealva aneurysm arising from the left coronary sinus, origin of the left anterior descending artery, stretched out left circumflex artery and the communication between the aneurysm and the left atrium.
Discussion

Most sinus of Valsalva aneurysms occur due to a congenital deficiency of elastic lamellae in the wall of the affected sinus, with separation of the media in the sinus from the media adjacent to the aortic valve annulus [6]. Progressive aneurysmal dilatation occurs due to long-standing high pressure and they eventually rupture [7]. An acquired sinus of Valsalva aneurysm is seen in conditions like tuberculosis, Behçet’s disease, infective endocarditis and trauma. Sinus of Valsalva aneurysms can present as subtle enlargement of an aneurysm sinus to tubular-shaped projection that extends from the apex or body of the sinus [8]. Sinus of Valsalva aneurysm may be associated with structural cardiac anomalies. Conal septal or perimembranous VSD is seen in 30–50% of patients with congenital sinus of Valsalva aneurysms [9,10]. Other anomalies are bicuspid aortic valve (10–20%), pulmonary stenosis, aortic coarctation, and atrial septal defect [4]. Modified Sakakibara classification for ruptured sinus of valsalva aneurysms is according to the site of the aneurysm rupture. The five types are as follows: type I, rupture into the right ventricle just beneath the pulmonary valve, type II, rupture into or just beneath the crista supraventricularis of the right ventricle; type III, rupture into the right atrium (type IIIa) or right ventricle (type IIIv) near or at the tricuspid annulus; type IV, rupture into the right atrium; and type V, other rare conditions, such as rupture into the left atrium, left ventricle, or pulmonary artery [11]. Our case is the rarest of all the Sakakibara types (Type V).

Imaging Techniques

Transthoracic echocardiography (TTE) delineates the sinus of Valsalva aneurysms if the acoustic window is adequate; however, there is need of transoesophageal echocardiography (TEE) in up to 25% of patients to depict the entire anatomy of the sinuses and its relationship to the surrounding structures [7]. TTE is advantageous as it is safe, cost-effective, and portable and acts as a first-line imaging modality in an emergency. Color Doppler imaging helps in identification of the fistulous connection.

Cardiovascular MRI helps in evaluation of cardiac morphology and is the standard technique which assesses the biventricular function. Quantification of the associated aortic regurgitation, shunt fraction in the setting of aneurysm rupture into the right heart chambers can be performed with ease. MRI has lower spatial resolution compared with CT but has higher soft-tissue contrast. MRI has contraindications like implanted ferromagnetic device, claustrophobia and has prolonged imaging time [12]. On MRI, a sinus of Valsalva aneurysm appears as a thin-walled out-pouching that is contiguous with the aortic root on sequential slices on black blood images. Functional impact on the valves and outflow tracts can also be assessed on bright-blood cine sequences, which depict dynamic changes in aneurysm morphology through the cardiac cycle. Flow jets as a result of aneurysm rupture may also be delineated.

ECG-gated MDCT can provide high-spatial-resolution images of the aortic root and in the non-emergent settings, MDCT offers advantages over echocardiography. Multiplanar reformations with 3D volume rendering provide excellent anatomic delineation and simultaneous evaluation of the coronary arteries. Retrospective ECG gating (continuous data acquisition through the cardiac cycle) provides valvular motion and ventricular function. However, it carries a significant radiation burden, especially for coverage of the entire thoracic aorta (15–20 mSv). With the introduction of the low-dose CT scanners with iterative reconstructions and tube current modulation,

Figure 3: Conventional angiography images showing contrast filled aneurysm from the left sinus of valsalva (A) and deployment of the Amplatzer PDA occluder device (B) and the check angiography (C) after the deployment showing non filling of the aneurysm from the left sinus of valsalva.

Figure 4: CT coronary angiography axial, sagittal and volume rendered images showing Amplatzer PDA occluder device across the origin of the aneurysm (A, C), decreased extravasation of the contrast (B) and volume rendered image (D) which shows the better filling of the coronary arteries and obliteration of the communication.

Figure 5: Schematic representation of the sinus of valsalva aneurysm (pre- and post- procedure). SVA- sinus of valsalva aneurysm, LV- left ventricle, Aorta, asterisk- left coronary sinus, AOD-Amplatzer occluder device.
dual source scanners, reduction in the radiation dose is possible. Sinus of valsalva aneurysm appears as a contrast filled out-pouching from the sinus and it may show a thrombus which appears as filling defect. It can also delineate the site of rupture. It also provides detailed anatomic depiction of surrounding cardiac structures and relationship of the coronary arteries. The high contrast resolution of CT also may make it possible to delineate the site of rupture for the aneurysms arising from the non-coronary sinus [9,10]. Rupture into the pericardial space is fatal and may occur from any sinus [4]. Clinical manifestations depend on the site and size of the fistulous connection. Large perforations may be visualized as a focal discontinuity in the aneurysm wall.

Arrhythmias: Atrioventricular node and His bundle dysfunction may be seen in aneurysms that expand or rupture into the interventricular septum. It is due to the direct pressure effects or adjacent low-grade inflammatory reaction [13]. Heart block and various degrees of bundle branch block have been also been reported [7].

Aortic Regurgitation: Sinus of Valsalva aneurysm is complicated by aortic regurgitation in 30–40% of the cases [4,9,10]. A lack of muscular support to the aortic valve annulus with altered hemodynamics of aortic root runoff leads to aortic regurgitation [5]. Aortic regurgitation volume may be calculated by comparison of left and right ventricular stroke volumes when there is no associated tricuspid or mitral regurgitation.

Outflow Tract Obstruction: RVOT and LVOT are obstructed by the sinus of Valsalva aneurysm of the right coronary sinus and left coronary sinus respectively [4,14].

Endocarditis: Sinus of valsalva aneurysm predispose to bacterial endocarditis [7]. Jet lesions can also develop on valves damaged by the fistulous stream, usually the tricuspid. Vegetations are visible on TEE and is considered a more accurate measure of assessment. This complication is an indication for urgent surgical repair [9].

Myocardial Ischemia: Disrupted aortic root an anatomy can cause kinking or compression of the coronary ostia with resultant myocardial ischemia and thrombus formation within a sinus of Valsalva aneurysm has the potential to embolize into the coronary arterial tree [15,16]. It is important to document the origins, course, and relationship of the proximal coronary arteries to the aneurysm as it determines the need for re-implantation at the time of surgery.

Aortogenic Embolism: A sinus of Valsalva aneurysm is a potential site of thrombus formation which may embolize to the systemic and cerebral circulation as well as the coronary circulation. Thrombus appears as a filling defect within the aneurysm on CT scans. On MRI, Chronic thrombus will have low to intermediate signal intensity on T1-weighted black-blood images and contrasts with the adjacent blood pool on bright-blood sequences. A thrombosed aneurysm must be distinguished from an intracardiac mass, such as myxoma. On MRI, A thrombosed aneurysm will appear contiguous with the aortic root on sequential bright-blood sequences. In cases of persisting uncertainty, delayed gadolinium enhancement can help because thrombus does not enhance centrally, whereas myxomas typically display a heterogeneous enhancement pattern [17].

Treatment Options

Both ruptured and unruptured sinus of Valsalva aneurysms are treated by surgical repair with a good ten year survival rate of 90% [5,9,10]. For symptomatic, nonruptured sinus of Valsalva aneurysms, the surgery is done if complicated by malignant arrhythmias, infective endocarditis, coronary artery compression, and RVOT obstruction [9]. The aorta and the chamber into which the aneurysm penetrates are accessed and resection of the aneurysmal sac is done, followed by the closure of the defect by pericardial or polyester patch or with sutures [18]. Reimplantation of the coronary artery is done if there is involvement of the coronary ostia by the aneurysm [4]. Aortic valve replacement and repair of any associated VSD may also be undertaken.

For nonruptured asymptomatic aneurysms, the optimal management strategy is less clear in the literature and an expectant management with serial imaging monitoring can be performed [9].

Percutaneous Trans-catheter closure of a sinus of Valsalva aneurysm is an excellent alternative to surgery with good results when there is no associated VSD or coronary ostial involvement. Few author groups have described the use of an occlusion device (Amplatzer Duct Occluder; AGA Medical Corp, Golden Valley, Minn) to successfully close right Valsalva sinus aneurysms ruptured into the right ventricle and right atrium, with no peri-procedural complications; all the four patients were asymptomatic at follow-up evaluations in their study [19-22]. Rao et al [23] described a case of successful trans-catheter coil occlusion of a right Valsalva sinus aneurysm that had ruptured into the right ventricle, with no reported periprocedural or late complications. Patients who have undergone sinus of Valsalva aneurysm repair require imaging follow-up as predisposition to recurrent aneurysm formation is present. Rittger, et al [24] reported a transcatheter closure of ruptured sinus of Valsalva aneurysma into the right ventricle with an Amplatzer Vascular Plug II and concluded that transcatheter closure is an excellent alternative to the surgery and the residual shunt can be present due to the inaccurate sizing of the plugs used for the procedure.

Our case is a unique case of aneurysm of left sinus of valsalva with rupture into left ventricle and left atrium which is the rarest variety of Sakakibara type V lesion. Percutaneous Trans-catheter treatment was performed to minimize the risk of future cardiovascular events and he underwent the closure of the lesion by Amplatzer PDA occluder device placement.

Conclusions

Sinus of valsalva aneurysm involves the right coronary sinus, followed by the noncoronary and left coronary sinuses. The ruptured and non-ruptured sinus of valsalva aneurysms vary widely in their clinical presentation. ECG-gated CT and MR imaging can provide excellent anatomic details. The mainstay of treatment is surgical repair with the surgical approach is by aortotomy or an approach from the chamber where the aneurysm terminates or a combination of the two [25]. Noninvasive percutaneous trans-catheter repair is an excellent treatment option when the patient has been diagnosed incidentally. Even though they are associated with fatal complications, the prognosis is excellent after the treatment; hence, it is important to make an accurate diagnosis.

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