Case Report
Operative, Echocardiographic and Angiocardiographic Details of an Adult with Subvalvular Aortic Aneurysm with Severe Aortic Regurgitation undergoing Patch Closure of the Aneurysm and Aortic Valve Replacement

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Introduction
Subvalvular aortic aneurysms are rare, poorly understood with variable etiologies and potentially lethal. As far as we are aware, and thus far there have been sporadic cases Subvalvular aortic aneurysms. The clinical presentation of Subvalvular aortic aneurysms varies, and the aneurysm is usually diagnosed by exclusion [1, 2, 3, 4, 5, 6, and 7]. Literature search till date revealed 28 cases of Subvalvular aortic aneurysms. Surgery remains the mainstay of treatment. Here-in we report an adult patient who presented with anginal pain and progressively increasing dyspnea with a diagnosis of Subvalvular aortic aneurysm and aortic regurgitation. The extreme rarity of the disease entity, the use of transesophageal echocardiography and computerized tomographic (CT) angiography for diagnosis, surgical details and a brief review of the published literature forms the basis of this communication.

Case Report
In January 2019, a 21 year-old was referred to our institute with recurrent episodes of anginal pain and progressively increasing symptoms of dyspnea [New York Heart Association class IV] of 2 years duration. She does not have a past history of rheumatic fever, tuberculosis or infective endocarditis or chest trauma. Notable clinical findings included a regular, hyper dynamic, high volume, collapsing pulse, blood pressure of 126/40mmHg, cardiomegaly, a systolic pericardial thrill, a grade 3/6 systolic and an early diastolic murmur loudest at the aortic area and mild hepatomegaly. Chest radiography revealed cardiomegaly of left ventricular type and mediastinal widening. Electrocardiogram showed sinus rhythm, diffuse T-wave inversions in anterolateral precordial leads with left ventricular hypertrophy. Transesophageal echocardiography showed wedge shaped echo free space between ascending aorta and left atrium [Figure 1].

Doppler echocardiogram demonstrated restrictive bi-directional flow into the aneurysm with a flow velocity of 4m/sec. Computerized tomographic angiography revealed an annular subaortic aneurysm in the atrioventricular groove underneath the right coronary cusp, measuring 2.0 cm transversely, 3.0 cm craniocaudally, and 2.5cm anteroposteriorly communicating with the left ventricular outflow.

Abstract
A 21 year-old woman with idiopathic subaortic aneurysm and severe aortic regurgitation was successfully treated by Dacron patch closure of the aneurysm and concomitant aortic valve replacement (AVR) and is reported for its rarity. The exact diagnosis is relatively easy to make by transesophageal echocardiography and/or computerized tomographic angiography, provided the possibility is entertained. Because of anginal pain and cardiac failure, surgical repair with AVR was deemed the best curative option to avoid further deterioration. Published reports detailing the diagnostic evaluation and surgical treatment of subaortic left ventricular annular aneurysms are also discussed. Transesophageal echocardiography is crucial in assessing the size of the ductus and confirming the adequacy of repair.

Keywords: Left Ventricular Aneurysms; Myocardial Ischemia; Subvalvular Aortic Aneurysm; Sudden Death
tract. There was mild extrinsic compression of the proximal left circumflex coronary artery by the aneurysm, however no calcification or atheromatous plaque was visualized [Figures 2A, 2B].

Figures 2A, 2B: Preoperative computerized three dimensional reconstructed volume rendered image showing Subvalvular aortic aneurysm underneath the right and left coronary cusp.

Intraoperative transesophageal color Doppler echocardiography confirmed the above findings. The operation was performed under cardiopulmonary bypass at 32° Celsius through angled venous cannulas into the superior and inferior caval veins and aortic cannulation. The left ventricle was vented through right superior pulmonary vein prior to cross clamp on a partially filled heart. After cross clamping the aorta, a transverse aortotomy was done in between stay sutures. Myocardial protection was achieved by intermittent selective ostial St. Thomas based cold hyperkalemic blood cardioplegia [1:4] and topical cardiac cooling using iced saline every 30 min.

The aortic valve exposure was facilitated by 3 commissural stay sutures. The aortic valve was excised. The orifice of the subaortic aneurysm was identified and repaired using a Dacron polyester patch [Bard® Savage® filamentous knitted polyester fabric, Bard Peripheral Vascular Inc., Tempe, AZ, USA] and interrupted pledgeted 4-0 polypropylene sutures [Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA]. The aortic valve was sized and replaced using a No. 21 mm St. Jude mechanical prosthesis [St. Jude Medical; St. Paul, MN, USA] and interrupted pledgeted 2-0 Ticron coated braided polyester sutures [M/s Covidien Santo Domingo, Dominican Republic, USA].

The aortic cross clamp and cardiopulmonary bypass time was 81 and 97 minutes respectively [Figures 3, 4].

Echocardiography did not reveal neither residual flow through the aneurysm or any paravalvular leak. At 6 months follow-up, she was asymptomatic, in NYHA functional class I and normal sinus rhythm. Post-operative CT angiography revealed complete interruption of blood flow through the sub aortic aneurysm [Figure 5].

Figures 3, 4, 5: Postoperative transesophageal two-dimensional color Doppler echocardiogram showing repaired Subvalvular aortic aneurysm with complete interruption of blood flow through the aneurysm and the replaced aortic valve. Postoperative transesophageal three-dimensional color Doppler echocardiogram showing repaired subaortic aneurysm (arrow) with complete interruption of blood flow through the aneurysm and absence of paravalvular leak. Postoperative computerized three-dimensional reconstructed volume rendered image showing repaired subannular aneurysm using a patch.

Discussion

Subvalvular aortic aneurysms are rare clinical entities of anatomical and surgical fascination. It was first described by JN Corvisart in 1812 [1]. It was an autopsy diagnosis, in whom the aneurysm was “almost as large as the heart itself”. It contained thrombus and communicated with the left ventricle through “an opening of small width, and whose contour was smooth and polished”. The first operation on aortic subvalvular aneurysm was performed in 1966 by Collins [2]. These aneurysms are usually smaller than the submitral ones and generally have a single chamber and a single small orifice. Despite their presumed congenital origin these aneurysms are slow to develop and are discovered only after they have grown to an average diameter of 3.7 cm (range 1.2 to 8.0 cm). Fewer than half contained thrombus. Rarely it is multilocular with double orifice [3, 4, 5, 6, 7, 8, and 9].

The orifice of the aneurysm is usually small, averaging 1.3 cm in diameter (range, 0.3 x 0.5 cm to 2.0 x 2.5 cm). The orifice is always contiguous to the aortic annulus on the ventricular side of the valve. Mostly, the orifice is located in the annulus fibrosus between the left aortic cusp and the anterior mitral leaflet. Other reported sites of origin are beneath the non-crownary and right coronary cusps and the
right and non-coronary cusp in the area of the membranous septum [3, 4, 5, 6, 7, 8, and 9]. Multiple subaortic aneurysms and combined submitral and subaortic aneurysms also have been reported.8 One-third of patients have associated aortic or subaortic anomalies, aneurysms of sinus of valsalva and aorta-left ventricular tunnel [3, 4, 5, 6, 7, 8, 9].

These aneurysms are less common the submitral aneurysms and are mostly evidenced in the literature as isolated case reports. Hence, the true incidence in the general population is unknown. They generally occur in young adults, although three have been reported in children. The average age at diagnosis is 28 years with a range of 6 to 46 years [4, 5, and 6], [10, 11, and 12]. Male sex predominates by a ratio of 2:1 among blacks. However, both sexes are equally affected among whites. Although initial reports were from Africa, only half of the total reported cases of subaortic aneurysms have been in blacks [4, 5, and 6], [10, 11, and 12]. Till 1993, 28 cases have been published; 26 of which were among the blacks. Twenty of these aneurysms were submitral and six occurred in the subaortic position [2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, and 13].

The genesis of subaortic aneurysm has been postulated to be congenital, resulting from a dehiscence of the fibromuscular union [8]. Its association with submitral aneurysm and aneurysm sinus of valsalva supports the concept of a congenital etiology [8, 9, 10, 11, 12, and 13]. The other suggested etiologies are post infective endocarditis, prosthetic valve endocarditis, cardiac tuberculosis, rheumatic carditis and syphilis. The causal effect of all these infections has remained speculative, as it may just reflect the high incidence of infections in endemic areas, rather than a causal mechanism [14, 15, 16, 17, 18, and 19].

The presenting symptoms are that of congestive cardiac failure with or without angina, myocardial infarction and sudden death [4, 5, 6, 7, 8, 9, 10, 11, 12, and 13], [17]. Its diagnosis has frequently been difficult on chest roentgenogram because of lack of characteristic findings but have been demonstrated by echocardiography and cardiac catheterization.

Transeosophageal echocardiogram is superior to transthoracic echocardiogram in delineating the neck and extent of the aneurysm [20]. CT angiogram and cardiac magnetic resonance imaging are standard investigative modalities to diagnose and characterize subaortic aneurysms and surgical planning [21, 22].

Subaortic aneurysms must be distinguished from sinus of Valsalva aneurysms. In our case, the sinus of Valsalva was normal on transeosophageal echocardiography and intraoperative inspection. Other developmental defects of conotruncus include aortico-left ventricular tunnel and sinus of valsalva aneurysms [8, 13]. Aortic regurgitation is commonly associated with subvalvular aortic aneurysms and may result from associated subaortic or aortic stenosis or from distortion and lack of support of the contiguous aortic cusp or from detachment of the aortic cusp [4, 5, 6, 7, 8, 9, 10, 11, 12, and 13].

Surgical repair is the treatment of choice for subaortic aneurysms, which are at risk of calcification, rupture and infective endocarditis, if left untreated [18]. Associated aortic, subaortic stenosis or aortic regurgitation demands surgical intervention in its own merit. Large aneurysms have been reported to compress the coronary arteries, causing angina, myocardial infarction and sudden death [4, 8, 9, 17, and 23] or to compress the conduction system causing heart blocks [24]. Our patient had coronary compression on CT angio and associated severe aortic regurgitation. In the published literature, obliteration of the orifice of the aneurysm was accomplished either by patch closure or by suture obliteration and plication. No recurrence or late complications have been reported. Like our case, concomitant AVR is also necessary in most patients [3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18, 19, and 20].

In cases of subvalvular aortic aneurysms with dilated aortic root, the traditional composite graft implantation technique especially in younger patients is the gold standard because of its durability [25]. Various modifications to this procedure including remodeling or re-implantation techniques have also been carried out. Although, there is evidence of excellent results following David's aortic valve-sparing technique and Yazhou's aortic root remodeling, many surgeons refrain from using this surgical alternative because it is technically demanding and time consuming [25, 26, and 27]. Furthermore, the incidence of aortic valve failure following valve-sparing procedure and bio-Bentall have not been adequately addressed in the published literature. There is currently no high level of evidence in favour of or against either of these procedures [25, 26, and 27].

Conclusions

The current case demonstrates that a subvalvular aortic aneurysm should be suspected in any young, non-hypertensive adult presenting with mediastinal widening on chest radiography with congestive cardiac failure or anginal pain. Surgical intervention in patients with aortic subannular aneurysms is often indicated because of coronary compression or associated aortic /subaortic stenosis or aortic regurgitation. Although the aneurysm can be treated by patch obliteration of the orifice, concomitant AVR is also necessary in patients with associated significant aortic valve disease. Transesophageal echocardiography plays a pivotal role in identifying, monitoring and planning surgical intervention in our patients.

In the absence of standardized treatment options, decisions to operate and type of surgery should be individualized in cases of dilated aortic root. We recommend an exhaustive search for other conotruncal anomalies like sinus of valsalva aneurysm, coarctation of the aorta and bicuspid aortic valve. The paucity of reports of subaortic aneurysms with aortic valvular disease has prompted us to report the case.

References


7. Yarom R, Grippel B. Aneurysm of interventricular septum with


