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UNRUPTURED LEFT CORONARY SINUS OF VALSALVA ANEURYSM WITH BICUSPID AORTIC VALVE STENOSIS AND LEFT VENTRICLAR INFERIOR WALL ISCHEMIA

Case Report

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ABSTRACT

Left coronary sinus of Valsalva aneurysm is a rare condition, and combination with aortic valve stenosis is very uncommon. The diagnosis by transthoracic echocardiography (TTE) could be difficult and can easily be interpreted as aortic root dilatation only. Even the computed tomography of the chest (CTscan) and transesophageal echocardiography (TEE) will not give a definite diagnosis unless we think of this pathology. We report a case of unruptured left coronary sinus of Valsalva aneurysm (ULSVA) in 39-year-old male patient combined with stenosis of bicuspid aortic valve and complete heart block. He has a permanent pacemaker (PPM) implanted and was operated successfully with aortic valve replacement (AVR) and direct closure of the ULSVA.

Keywords: Sinus of Valsalva aneurysm, Left coronary sinus, Bicuspid aortic valve stenosis, Myocardial ischaemia.

INTRODUCTION

Sinus of Valsalva aneurysm (SVA) is a localized thinning and weakness of the aortic sinus wall presenting with bulging just above the annulus of the leaflet hinge. It could be congenital due to the absence of the elastic and muscular tissue of the sinus wall (1), with entirely intracardiac course (1), or acquired secondary to medionecrosis, syphilis, atherosclerosis, bacterial endocarditis, and penetrating injury (1). The acquired SVA has more diffuse course involving most of the sinus or multiple sinuses and often the ascending aorta, and projecting therefore into the pericardium outside of the heart. Generally, in case of bicuspid aortic valve (BAV) the genesis of the SVA and aortic dilatation is not fully elucidated, and most probably is a combination of congenital aortic fragility and accelerated degeneration of the aortic media (9), with consequent changes because of hemodynamic burdens caused by aortic stenosis (AS) or aortic regurgitation (AR). The maximum dilatation of the aorta in case of BAV alone is at the levels of sinotubular junction (STJ) and ascending aorta (AA), and less at the aortic root level, (9) which suggests the congenital component of the development of SVA in our case - the maximum dilatation is above the aortic valve in one sinus of Valsalva only.

It presents clinically during the 3rd or 4th decade of life, more often after perforation into a cardiac chamber causing a fistula, obstruction of the right or left ventricular outflow tract, compressing the coronary arteries and surrounding structures.

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CASE REPORT

A 39-year-old man, second admition to Chest Diseases Hospital in Kuwait, because of aortic valve stenosis (AVS) and aortic root enlargement, with complains of progressing shortness of breath, decreased exercise tolerance for the last few months - NYHA class III on admission and atypical chest pain for the last four years. The physical examination revealed a grade 3/6 systolic murmur all over the precordium with radiation to the carotids, blood pressure of 140/80 mmHg, and the pulse rate 70 beats/min (DDDR pacer mode 60 - 120 beats/min). Chest X-Ray showed mild cardiomegaly with dilated aortic arch. TTE showed heavily calcified bicuspid aortic valve, moderate aortic stenosis (AS) with peak transvalvular gradient of 49 mmHg, mean gradient 28 mmHg, mild posterior dilatation of ascending aorta (AA) 49 mm. There is no AR, and the other valves were normal. Left ventricle ejection fraction (EF) was 60%. The pa-

tient underwent coronary angiogram which confirmed moderate AS. He is a known case of congenital bicuspid aortic valve and incomplete right bundle branch block (RBBB). A permanent pacemaker (PPM) has been implanted 4 years ago because of complete atrio-ventricular block (A-V block). Dipyridamol stress myocardial perfusion scintigraphy done three years ago showed a small fixed perfusion defect in the apex and mild stress induced ischemia in the inferior wall of the myocardium.

His symptoms has deteriorated rapidly over the last 3 months, so the catheterization was repeated and revealed left ventricle EF decreased to 30%. Aortic root angiography showed bicuspid valve with calcified anterior leaflet, large aneurysm of the posterior, left coronary sinus of Valsalva and the diameter of the aortic root measured 60 mm. Ascending aorta is dilated, but significantly smaller than the root (Fig.1).

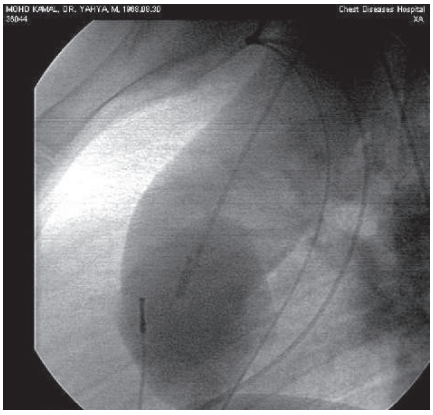

Fig. 1. Left Coronary Sinus of Valsalva Aneurysm

His coronary arteries are normal, with a very short (1-2 mm) left main stem arising at the upper part of the posterior coronary sinus. The circumflex branches are following the contour of the LSVA, and the first Obtus marginal branch (OM1) supplies the LV inferior wall towards the apex (Fig.2).

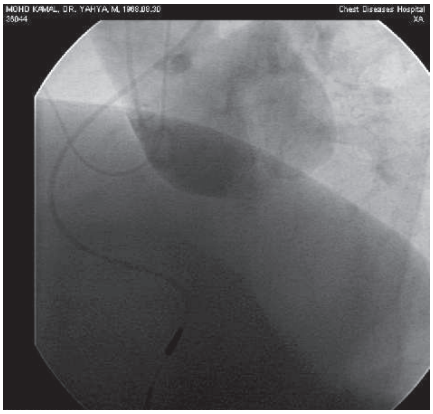

Fig. 2. Left Coronary Sinus of Valsalva Aneurysm

The right coronary artery is small, non dominant, arising from the anterior sinus of Valsalva. There are no significant changes in the size of cardiac chambers and trans-valvular gradient compared with previous examination. Contrast enhanced CT scan demonstrated aortic root enlargement up to 60 mm, but failed to distinguish the dilatation of the aortic root from much less dilated ascending aorta, and the difference between the anterior and posterior coronary sinuses. Only the TEE done into the operating room after the induction of anesthesia rouse the suspicion of left sinus of Valsalva aneurysm (LSVA).

The patient was operated on through a median sternotomy with mild hypothermic cardiopulmonary bypass. The dimeter of the ascending aorta measured by TEE was about 47 mm, which was considered as an acceptable size for a man with 139 kg of weight and 198 cm height. The aortic valve was approached through an oblique aortotomy. The calcified anterior leaflet and completely free of calcium and mildly fibrotic posterior leaflet of this bicuspid valve were excised, and the calcium debris were removed from the contiguous interventricular septum. Left sinus of Valsalva aneurysm was situated above the valve, with the left coronary ostium arising at the upper part of the sinus, approximately 3 cm above the valve (Fig.2). There was no significant dilatation of the anterior sinus of Valsalva. The direct closure of the LSVA by plication and aortic valve replacement with a # 25 St.Jude mechanical valve were performed and the aortotomy was closed. Postoperative TEE demonstrated aortic root diameter reduced to 49 mm.

DISCUSSION

The incidence of SVA or fistula is between 0.14% and 0.96%, with 80% male predominance (3). The combination of unruptured left sinus of Valsalva aneurysm, bicuspid aortic valve with AS and complete A-V block obviously is very rare and we couldn't find in "PubMed - index for MEDLINE" such a case described and published in the last 15 years.

In 70 to 90% of cases the SVA arises from the right coronary sinus, and very rare from the left. In the typical case it is combined with aortic regurgitation (AR), eventually with perforation into right atrium or ventricle. When the unruptured SVA is arising from the left coronary sinus, the diagnosis by TTE could be very difficult and only trans-esophageal echocardiography (TEE) or aortic root angiography are informative enough. The conventional CT scan probably is not a reliable method for the diagnosis of SVA - the differentiation between dilatation of the sinus of Valsalva alone from whole aortic root dilatation is difficult or impossible. In the case we reported, the pathogenesis is most probably related to both congenital abnormalities of the aortic wall and bicuspid aortic valve, and acquired changes of the sinus of Valsalva caused by AS and dilated coronary sinus as well. Calcification of the anterior leaflet was diagnosed long time before clinical presentation. Because of the bicuspid aortic valve it was very difficult to guess whether the complete A-V block is developed as a consequence of the calcium penetration into the

interventricular septum, or as the result of the pressure created on the displaced and dilated posterior coronary sinus (1). The complaint of chest pain and the sings of LV inferior wall and apical ischemia discovered by radioisotope examination might be a result of the anatomical relation between the left coronary sinus and the proximal left coronary artery (7). We think that the left main stem and the circumflex branches which were lying behind the LSVA are compressed and stretched during the heart diastole, although the coronary arteries were normal on the angiography. Repeated TTE and CT scan did not achieve the diagnosis of the ULSVA pre operatively, and the main indication for the surgical treatment of this patient was the clinically symptomatic aortic stenosis. Aortic valve replacement and closure of the ULSVA by plication were effective to reduce the size of aortic root significantly. Patient should be followed up regarding the possibility of further aortic root and ascending aortic dilatation, a tendency well described in patients with bicuspid aortic valve, and for sings and symptoms of myocardial ischemia. The only reliable methods for the assessing the morphology of the posterior coronary sinus and sinotubular junction are aortic root angiography and TEE. These can distinguish between LSVA from whole aortic root dilatation, which is important for the operative strategy. CT angioscopy is the new methods for diagnosis of aortic root pathology and eventually could be used successfully.

CONCLUSION

During the 4th and 5th decade of life there is clear tendency towards complications of the sinus of Valsalva aneurysms. If diagnosed, perforation in cardiac chambers or structures, or compression of surrounding structures, or even non complicated but large SVA should be treated surgically to prevent further complications like heart failure, endocarditis, heart block, bleeding etc. The proper surgical treatment, with or without aortic valve replacement or repair, has reasonable low mortality rate and good long-term results. Because of the congenital aortic wall weakness, follow up by TTE, and TEE if needed, is advisable.

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