Aneurysms of sinus of Valsalva eroding into the interventricular septum: etiopathology and surgical considerations

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Received 24 March 1997; received in revised form 5 August 1997; accepted 1 September 1997

Abstract

Objective: To evaluate and discuss etiopathology, clinical manifestations and surgical outcome of a rare subset of unruptured aneurysm of the sinus of Valsalva which erodes into the interventricular septum. Methods: Between 1989 and 1995, seven cases of unruptured aneurysm of the sinus of Valsalva eroding into the interventricular septum underwent surgical correction at the King Edward VII Memorial Hospital, Bombay. The origin of all these aneurysms was from the right coronary sinus. The mean age of presentation was 31 years. All patients were male. Calcification of the aneurysm was seen in three. Three patients presented without aortic regurgitation; all had complete heart block. Four patients presented with aortic regurgitation and in addition, two had complete heart block. Preoperative left ventricular function was poor in patients with aortic regurgitation (Ejection fraction range: 30–42%), when compared to those without aortic regurgitation (Ejection fraction range: 48–52%). Of those without aortic regurgitation at initial presentation, one patient developed progressive aortic regurgitation after 3 years requiring surgery. While two other patients were operated at earliest for closure of aneurysm, even in the absence of aortic regurgitation. All those with aortic regurgitation required surgery for aortic valve replacement and closure of aneurysm. Aneurysm was closed by direct suturing of the ostium in two patients and by patch closure in five patients. Permanent pacemaker was implanted in five patients. Result: There was no operative death. Patients who underwent aortic valve replacement required postoperative inotropic support. Two patients, who underwent surgery in absence of aortic regurgitation, remain free of aortic regurgitation at the end of 36 and 42 months of follow-up. One of the patients with calcific aneurysmal sac underwent successful re-replacement of the aortic valve for paravalvar leak after a 2 year interval. Conclusion: Unruptured aneurysm of the sinus of Valsalva eroding into the interventricular septum should be operated at the earliest, which makes surgery simple and prevents development of complications such as aortic regurgitation and heart block. © 1997 Elsevier Science B.V.

Keywords: Aneurysm; Sinus of Valsalva; Aortic regurgitation; Heart block; Cardiac calcification

1. Introduction

Aneurysms of sinus of Valsalva (ASOV) are usually congenital in origin, mostly arising from the right coronary sinus or the adjacent half of the non-coronary sinus of Valsalva [1–4]. Unruptured ASOV can cause significant anatomic and physiological derangement and may present with evidence of right ventricular outflow tract obstruction, aortic insufficiency, conduction abnormalities or coronary artery compression [5–7]. Dissection of the interventricular septum (IVS) is a rare complication of ASOV. In this report, we analysed seven cases presented during the last 16 years, with special reference to noninvasive methods of diagnosis of this rare clinical condition.
2. Material and methods

During the period between 1979 and 1995, a total of 47 cases of ASOV underwent surgical correction at the K.E.M Hospital, Bombay. These patients constituted 0.2% of patients who underwent cardiac surgery utilizing cardiopulmonary bypass (CPB) during this period. Of these, seven patients were diagnosed as unruptured ASOV extending into IVS. Diagnosis was confirmed in all the patients during surgery. Interestingly, all seven cases occurred between 1989 to 1995. All seven were male patients, ranging in age from 12 to 56 years (mean age 31 years). Aortic regurgitation (AR) was present in four patients from (Grade IV/IV in three and III/IV in one). There was no associated ventricular septal defect (VSD).

Three patients without AR presented with giddiness secondary to CHB and the remaining four patients presented with clinical evidence of AR; of these, two also had CHB. All patients complained of gradual onset of dyspnoea and were in NYHA functional class II (3/7 of those without AR) or III (4/7 of those with AR). There was no history of infective endocarditis, angina or thromboembolic episodes in any of them. One of the patients without AR at initial presentation, developed progressively worsening AR with increase in the size of the aneurysm and worsening of left ventricular function within 3 years, requiring closure of aneurysm with aortic valve replacement.

On roentgenogram, mild to moderate degree of cardiomegaly was evident in patients with aortic regurgitation. A calcific contour in the area of aortic root and margins of the aneurysm was evident in three patients (Fig. 1). Two-dimensional echocardiography and Doppler performed with a Hewlett-Packard SONOS 1000 ultrasound system (Hewlett-Packard, Andover, MA) revealed an aneurysm arising from the right coronary sinus extending into the IVS. Size of the aneurysm varied from 2.4 × 3.0 cm to 4.2 × 7.2 cm. Continuation of echofree space within the IVS with right coronary sinus was evident in all (Fig. 2A), while the characteristic diastolic expansion with systolic emptying was demonstrated in only those cases without calcification (Fig. 2BC). There was evidence of AR of varying severity in four patients. Left ventricular function was significantly lower in patients with AR (Ejection fraction range 30–42%) when compared with those without AR (Ejection fraction range 48–52%). One of the patients without AR at initial presentation developed progressive AR and deterioration of left ventricular ejection fraction from 48 to 34% within 3 years.

Angiography was performed in two cases, in one who presented in 1989 when we did not have color Doppler facility and in another (age 56 years) to rule out coronary artery disease. The ascending aortic angiogram showed a large pulsatile aneurysm in one (Fig. 3AB), while in the other it was found to be non pulsatile, probably due to the presence of calcification.

2.1. Operative findings

All repairs were performed using standard CPB with moderate hypothermia and multiple cold crystalloid cardioplegia for myocardial preservation. After cross clamping the aorta through a low transverse aortotomy, aortic valve was inspected for presence of vegetations, perforation and nature of the cusps. Orifice of the aneurysm was then identified (which ranged from 5 to 20 mm) (Fig. 4): all were in relation to mid right coronary sinus. Calcification was obvious in one involving the entire aneurysm. In two patients, surgery was done in absence of AR. In these patients, aneurysmal opening was small (5 and 6 mm). Direct closure of the opening of the aneurysm was performed with polypropylene (4-0) sutures1 reinforced with teflon pledgets. Aortic cusps were of good quality with no evidence of AR and hence, aortic valve was not replaced or repaired. In the remaining five patients, the aorta was dilated, aortic cusps, mainly the right coronary cusp, were distorted with poor approximation. The

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Fig. 1. Fluoroscopy. Arrows point to areas of calcification in the aneurysmal wall which extends into the interventricular septum.

1 Ethicon, Edinburgh, U.K.
aneurysmal sac opening was large (16 to 20 mm) and three patients had a calcific aneurysmal sac. Hence, patch repair of the aneurysm was carried out, followed by aortic valve replacement in all these patients. Two of these patients had calcification of the aneurysm. Mechanical prostheses were used in all these patients: 23 mm Starr Edward prosthesis in three patients, 23 mm Chitra in one and 23 mm Carbomedic prosthesis in one patient. Interrupted Ethibond (2-0) sutures were used for implantation of the valve. Aortotomy was closed. All patients with AR required high ionotropic support to come off bypass (dopamine 7.5–10 μg/kg per min). Epicardial leads of permanent pacemaker were implanted in four patients, as one patient with CHB already had a transvenous pacemaker implanted before surgery. Important patient characteristics are summarised in Table 1.

3. Results

There was no operative mortality. All patients with associated AR were maintained on ionotropic support for 36–48 h. Patients were followed up for 3 months to 7 years with clinical, radiological and echocardiographic examinations (mean follow-up of 3 years and 2 months). At the time of last follow-up all were in NYHA class I–II. Of the seven, two patients without heart block continue to be in sinus rhythm. In those with a permanent pacemaker, there was no improvement in the status of CHB. Roentgenogram showed

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2 Baxter Edwards A G, Switzerland.
3 TTK Pharma, India
4 Carbomedics, Texas.
normalisation of heart size in all four patients with preoperative AR. Echocardiography revealed good prosthetic valve function with reduction in left ventricular dimensions. Aneurysmal sacs were nearly obliterated in most patients, except in three patients with calcific sacs. One of the patients underwent successful re-aortic valve replacement after 2 years of initial surgery for a significant paravalvar leak, detected clinically and confirmed at echocardiography and color Doppler studies. This patient had a calcific aneurysmal sac and the leak was in the region of the calcific margin of the aneurysm.

4. Discussion

ASOV is an uncommon anomaly, attributed to deficiency or absence of media in the aortic wall [8,9]. Orientals are apparently more prone to develop this pathology [10,11]. A total of 20% of ASOV have no perforation and are identified only by chance at surgery or at autopsy or if a complication supervenes, such as obstruction of the right ventricular outflow tract, aortic insufficiency, conduction abnormalities, ventricular tachycardia, coronary artery compression, infective endocarditis, left ventricular outflow obstruction or it can be a source of an embolus [5,7,8,12–16]. With advent of echocardiography, unruptured ASOV are diagnosed with increasing frequency [11]. The extension of an unruptured ASOV into the IVS is extremely rare [17–23].

All our patients were males and the sinus of origin was the right coronary sinus, which is consistent with the previously reported cases. [17–23]. Age of the patients varied from 12 to 56 years (mean age of 31 years).

The mechanism for the septal extension has been ascribed to simple progression, intraseptal rupture with hematoma and subsequent psuedoaneurysm and bacterial endocarditis [24]. Various etiologies have been implicated such as congenital, bacterial endocarditis, syphilis, trauma, Marfan’s syndrome, dissection of aorta, rheumatoid arthritis and atherosclerotic degeneration [24]. A specific etiology could not be detected in any of our patients.

A common feature reported with septal extension of the aneurysm is the presence of atrioventricular conduction disturbances [17–23]. The mechanism for atrioventricular block is easily explained by extension of the ASOV to the vicinity of the conduction tissue [22]. Most frequent of the conduction disturbances is CHB (in 12 patients), followed by left bundle branch block (in 5 patients) and right bundle branch block (in 1 patient). In a few cases, evolution from trifascicular block to complete heart block was observed [22]. In our series, complete heart block was seen in five patients. The remaining two patients were in sinus rhythm. CHB needs pacemaker implantation.

Aortic regurgitation, minimal to severe, is present in the majority of cases of ASOV, whether ruptured or not [17–23]. The mechanism, in the presence of an aneurysm extending into the IVS, is lack of septal support to the leaflet which seems to result in progressive AR. This is supported by the fact that in one of our patients, AR developed later, with a corresponding increase in the aneurysmal size requiring aortic valve replacement. Hence with early surgery, it may be possible to approximate the edges of the aneurysmal orifice, thus reinforcing the aortic annulus and supporting the cusp [17,19]. Further, those with AR required ionotropic support for a longer duration, reflecting on their relatively poor left ventricular function which could be due to mechanical interference with the septal motion by the aneurysm, associated AR and in some, extension of calcification into the myocardium [9].

Calcification of ASOV is rare. Of the seven patients, three had calcification which extended along the annulus. We found only two previous reports in the
literature [19,22,25,26]. The favorable consequences of calcification are less ventricular dyskinesia due to rigidity of calcific wall of aneurysm and halting of further progression of the aneurysm. The unfavorable consequences are that it renders direct suturing of the aneurysmal orifice impossible, increases chances of paravalvar leak following aortic valve and may contribute to the poor left ventricular function [22].

Diagnosis of unruptured ASOV has evolved from postmortem findings in 1956 by Lee et al., to preoperative diagnosis [20]. The first preoperative diagnosis of this condition was reported by Engel et al., in 1980 [20]. Before the advent of echocardiography, cardiac catheterisation was the principal diagnostic method [18,22]. It served to establish diagnosis by demonstrating continuity between the aneurysm and the aortic sinus and could demonstrate filling of the sac, its pulsations (Fig. 3A B).

Lewis and Agathangelou demonstrated on echocardiography, the continuity between the echo free space with the upper septum and the aortic root at the level of the right coronary sinus [14] (Fig. 2A). Hands et al., further described the characteristic systolic emptying and diastolic expansion in such cases [21] (Fig. 2BC). The second feature may be absent in severely calcific aneurysms due to rigid walls. The conditions which can be confused with this are ventricular septal cysts and septal aneurysms. Transesophageal echocardiography has also been found in diagnosing this condition and can detect intra-aneurysmal pathologies like thrombus and vegetations [16,27]. Recent interest has developed into Cine phase contrast Magnetic Resonance (MR) detection of ruptured ASOV [24,28]. The presence of a permanent pacemaker in many of these patients limits its use in this condition.

Although the implications and natural history of an unruptured ASOV are not clear, surgery forms the only definitive treatment and has prophylactic value in preventing complications such as AR and development of conduction defects. Conservative treatment is possible if AR is absent or mild and if the aneurysm is not increasing further [14,18]. During surgery, resection of the pouch is unnecessary and may damage the aortic valve and IVS. Closure of the mouth of aneurysm with either direct sutures or with a patch is a simple and effective treatment when operated upon early. Aortic valve repair or replacement is necessary when operated at a late stage, as seen in our patients.
We conclude that unruptured ASOV dissecting into IVS is a distinct entity, with respect to its presentation, complications and treatment. Transthoracic echocardiography is adequate for diagnosis and post-operative follow-up. Early surgery prevents development and progression of AR and may also prevent development of conduction abnormalities. AR is the major indication for surgery in these patients and requires aortic valve replacement in most. Prognosis is good, if operated at an early stage.

Acknowledgements

We thank our Dean, Dr Pragnya Pai of Seth G.S. Medical College and King Edward VII Memorial Hospital, Parel, Bombay for allowing us to publish this data.

References


