Case report - Congenital

Surgical repair of multiple unruptured aneurysms of sinus of Valsalva

Srikrishna Modugula Reddy, Akshay Kumar Bisoi*, Pranav Sharma, Shambunath Das

Cardiothoracic Centre, All India Institute of Medical Sciences, New Delhi, India

Received 27 March 2009; received in revised form 16 June 2009; accepted 18 June 2009

Abstract

Unruptured aneurysm of sinus of Valsalva (ASV) is a rare congenital anomaly. We describe a case of multiple unruptured ASV involving right and left aortic sinuses causing congestive cardiac failure in a 16-year-old boy who underwent successful surgical repair.

Keywords: Aneurysm; Sinus of Valsalva; Aortic root

1. Introduction

Unruptured aneurysm of sinus of Valsalva (ASV) is a rare congenital anomaly. We describe a case of multiple unruptured ASV involving right and left aortic sinuses nearer to origin of coronary ostia with extensive dissection into the interventricular septum (IVS) and left ventricular (LV) wall causing congestive cardiac failure in a 16-year-old boy who underwent successful surgical repair.

2. Case report

A 16-year-old boy presented to our institute with symptoms of progressively increasing dyspnea and palpitations of one year duration. Clinical examination revealed a regular pulse rate of 105/min, blood pressure of 110/60 mmHg, a diastolic thrill over precordium, a loud holodiastolic murmur over left third intercostal space and few basal pulmonary crepts. A 12-lead electrocardiogram showed trifascicular block with intermittent ventricular ectopics. Chest X-ray revealed cardiomegaly with features suggestive of pulmonary venous hypertension. Blood biochemical and hematological parameters were within normal range.

Transthoracic two-dimensional echocardiography (Fig. 1) revealed a partly calcified, multicycstic echo-free space originating from the dilated and deformed right aortic sinus, extensively extending into the IVS up to the apex and all around the LV lateral wall with a significant diastolic regurgitation into the false cavity without any valvular lesion. Aneurysm arising from the left aortic sinus contained a large layered thrombus. In addition, there was mild mitral regurgitation and moderate ventricular dysfunction.

Intra-operative findings (Fig. 2a) included normal aortic valve leaflets and two unruptured ASV. One was arising from the right aortic sinus close to right coronary artery ostium, burrowed into the IVS up to the apex and all around the LV lateral wall without any ventricular connection and contained only blood. Another was arising from the left aortic sinus close to left coronary ostium, burrowed into aortomitral curtain mostly extending externally and posteriorly above the left atrial roof and contained laminated clots. Surgery was performed through a median sternotomy incision. Under cardiopulmonary bypass and moderate hypothermia, the aorta was cross-clamped and an oblique aortotomy was performed. Antegrade cold blood cardioplegia was delivered through the coronary ostia. Following assessment of aortic valve, mouth of the aneurysms were identified, aneurysm cavity was filled with normal saline to avoid air entrapment and subsequent air embolism and repaired by exclusion of the aneurysm sac and closure of the sinus defects using expanded polytetrafluoroethylene patch (Fig. 2b and c). Aortic valve was reassessed and aortotomy closed. Patient was weaned off from cardiopulmonary bypass in a standard manner. Intraoperative trans-esophageal echocardiography revealed normal aortic and mitral valves without any evidence of stenosis or regurgitation.

Postoperative course was uneventful. A permanent pacemaker was implanted prophylactically on day 12 following surgery in view of the persistent features of trifascicular block on 24 h Holter monitoring. At nine months follow-up, the patient remains asymptomatic and doing well. Two-dimensional trans-thoracic echocardiography revealed substantial reduction in the size of cystic spaces within the ventricular septum without any flow, normal aortic and mitral valves and normal LV function.

*Corresponding author. Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, 110029, India. Tel.: +91-11-26594835; fax: +91-11-26588663.
E-mail address: akshaykbisoi@gmail.com (A.K. Bisoi).
© 2009 Published by European Association for Cardio-Thoracic Surgery.
3. Discussion

ASV [1] is a rare defect of the aortic root and accounts for 0.14–1.5% of the cardiac surgical load, the incidence being higher in patients of Asian origin. Further, an unruptured ASV that dissects into the IVS is an extremely rare entity [2]. An even greater rarity is the presence of multiple unruptured aneurysms in the same patient, and to date only 14 cases have been reported in the English language literature. The causes of ASV include congenital, cystic medionecrosis, atherosclerosis, connective tissue disorders, infections and trauma [3].

Unruptured ASV is asymptomatic usually and often found incidentally. However, the condition can manifest by distortion or compression of adjacent structures as aortic insufficiency, right ventricular outflow tract obstruction, tricuspid stenosis and regurgitation, myocardial ischemia or infarction, conduction disturbances with septal penetration, mediastinal mass, infective endocarditis, thromboembolism and rupture. The diagnostic modalities include trans-thoracic and trans-esophageal echocardiography, computerized tomography, angiography, cardiac catheterization and magnetic resonance imaging [3].

Surgical repair of unruptured ASV should be advised in symptomatic cases, in cases of rupture, and in incidentally discovered cases at time of surgery for other cardiac pathology unless specific contraindications exist. These aneurysms are probably best repaired by exclusion of aneurysm sac and closure of sinus defect using expanded polytetrafluoroethylene patch or pericardial patch [1]. The use of expanded polytetrafluoroethylene patch is preferred over pericardial patch closure or direct closure of the defect to prevent the possibility of aortic valve leaflet deformity and/or coronary ostial narrowing. Surgical repair may involve adjuvant procedures like aortic valve sparing procedures or aortic valve or aortic root replacement in cases of associated aortic regurgitation or aortic root distortion. As the surgical repair offers no guarantee against the fatal conduction abnormalities, prophylactic permanent pacemaker implantation was preferred in patients having extensive ventricular septal extension as seen in our case [1].

The presence of unique combination of findings namely multiple unruptured aneurysms involving right and left aortic sinuses nearer to origin of coronary ostia, extensive dissection into ventricular septum and LV wall, no true valvular aortic regurgitation despite severe distortion of sinuses and no complete heart block despite extensive involvement of ventricular septum in our patient made this a very unusual case that was successfully managed.

In conclusion, unruptured ASV is a distinct and a rare clinical entity with respect to its presentation, complications and treatment. Once diagnosed, early surgical repair is recommended.
References

