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Case Report

A case of unruptured right sinus of Valsalva with intraventricular septal dissection and complete heart block

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ABSTRACT

Unruptured right sinus of Valsalva causing intraventricular septal dissection and presenting as complete heart block is an extremely rare entity. We report a 38 years old male patient presenting with an episode of syncope. Electrocardiogram revealed complete heart block. Echocardiography and aortic root angiogram showed an aneurysm from right sinus of Valsalva dissecting the intraventricular septum. Patient refused operation and only permanent pacemaker implantation was done for complete heart block.

1. Introduction

Aneurysm of sinus of Valsalva accounts for 1% of congenital anomalies of heart and circulation.[1] In most cases unruptured sinus of Valsalva aneurysm is clinically silent, diagnosed incidentally. However unruptured congenital sinus of Valsalva aneurysm may cause symptoms by obstructing right ventricular outflow tract, distort aortic valve causing incompetence, compress coronaries causing myocardial ischemia and rarely conduction disturbances or even complete heart block by compressing conduction system. In this case report we describe a case of unruptured aneurysm of right sinus of Valsalva with extension into intraventricular septum causing complete heart block.

2. Case

A 38 years old man presented with history of acute onset giddiness since last 10 h with no previous history suggestive of cardiac disease. On examination pulse was 44 per minute, regular of good volume. Blood pressure was 122/74 mm Hg. Jugular venous pulse showed prominent a-wave. A harsh ejection systolic murmur of Grade III was audible maximal in 3rd left intercostal space along the left sternal border. A decrescendo murmur of Grade II was audible maximal in 2nd left intercostal space along left sternal border.

Electrocardiogram revealed atria-ventricular dissociation with atrial rate of 120/minute and ventricular rate of 42/minute. Chest radiograph was unremarkable. Transthoracic two

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dimensional echocardiography demonstrated an aneurysm arising from right sinus of Valsalva and dissecting the intraventricular septum (Figs. 1 and 2). An aneurysmal clot was seen in parasternal short axis view (Fig. 3). Colour Doppler echocardiography showed turbulent flow from right sinus to aneurysm with orifice diameter of 12 mm. Also demonstrated was mild aortic regurgitation with normal left ventricular function and no features of infective endocarditis.

The echocardiographic findings were further confirmed by aortic root aortogram showing unruptured aneurysm from right sinus of Valsalva dissecting into intraventricular septum with aortic regurgitation (Fig. 4 A and B).

Surgical repair was offered to patient; but patient refused, and only permanent pacemaker was implanted.

3. Discussion

Aneurysm of sinus of Valsalva accounts for 1% of congenital anomalies of heart and circulation. The fundamental histologic fault responsible is the discontinuity of elastic layer in the aortic media at the juncture between the ascending aorta and the aortic valve annulus.[2,3] On one hand most of the unruptured sinus of Valsalva is diagnosed incidentally and on the other the patient may present with sudden death resulting from rupture to pericardium and occasionally complete heart block.

90%–95% originate in the right or non-coronary sinus and project into the right ventricle and right atrium, and less than 5% originate in the left coronary sinus. Those that arise in the non-coronary sinus almost always rupture into the right atrium and those that arises in right coronary sinus ruptures into the right ventricle or occasionally into right atrium.

Extension to intraventricular septum is rare and is ascribed to the proximity of right sinus of Valsalva especially the middle part. Possible theory put forward to explain the pathologic mechanism of intraventricular extension causing complete heart block is the direct pressure by the expanding aneurysm to the conducting tissue. However Lee and colleagues[4] found evidence of hemorrhage and low grade inflammatory changes in the conduction tissue and held these inflammatory changes responsible for it. This was further supported by the fact that the block reversed with corticosteroid therapy.

Beside aortic regurgitation and heart block unruptured sinus of Valsalva may present as a source of cerebral embolism[5] or even a focus for infective endocarditis.[6]

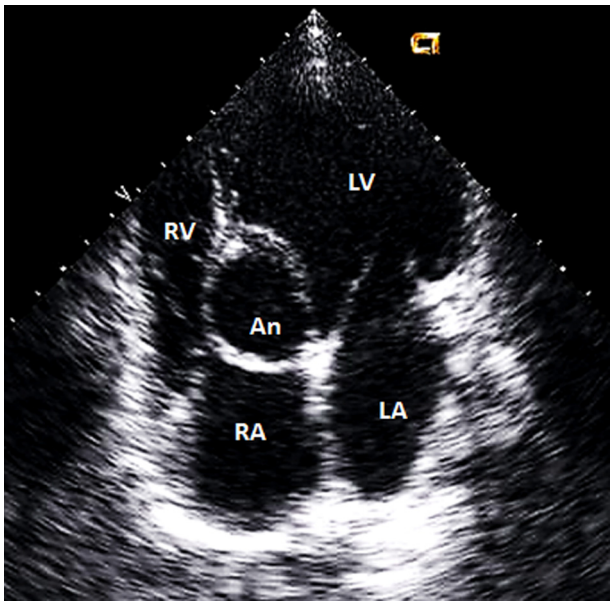

Fig. 2 – Transthoracic two dimensional echocardiography; apical four chamber view showing an unruptured sinus of Valsalva dissecting the intraventricular septum. An: sinus of Valsalva aneurysm, LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

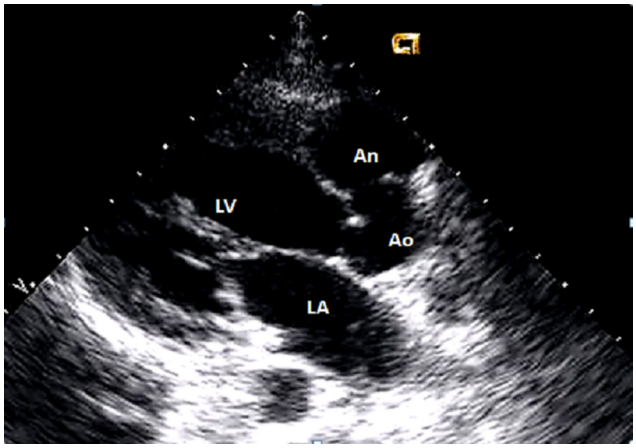

Fig. 1 – Transthoracic two dimensional echocardiography; parasternal long axis view revealing an unruptured sinus of Valsalva aneurysm arising from right coronary sinus. An: sinus of Valsalva aneurysm, Ao: aorta, LA: left atrium, LV: left ventricle.

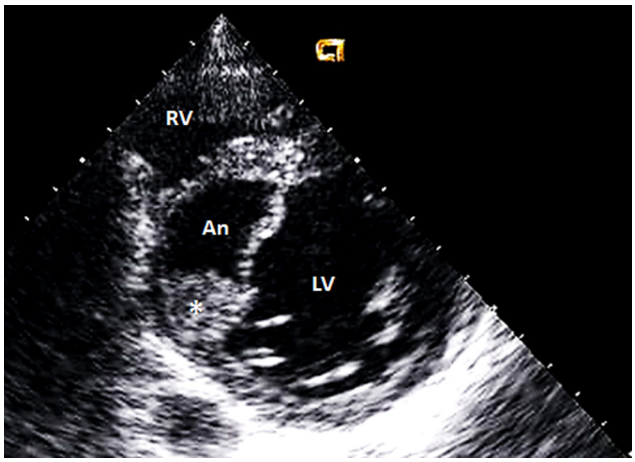

Fig. 3 – Transthoracic two dimensional echocardiography; parasternal short axis view at mitral valve level revealing unruptured sinus of Valsalva dissecting intraventricular septum and containing aneurysmal clot. An: aneurysm of sinus of Valsalva, LV: left ventricle, RV: right ventricle, *: Aneurysmal clot.