Case Report

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A Case of Right Sinus of Valsalva Rupture with Dissection into Interventricular Septum Causing Left Ventricular Outflow Tract Obstruction

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Sinus of Valsalva aneurysm (SVA) is an uncommon anomaly of the aorta. Rupture of SVA often precipitates dramatic clinical complications, including heart failures. Right SVAs are the most common type, and when they rupture, they usually rupture into the right ventricle or right atrium. Rupture into left ventricle or interventricular septum is rare. Herein, we report a case of right SVA rupture with dissection into interventricular septum, which produced significant left ventricular outflow tract obstruction and aortic regurgitation. The case was successfully treated by surgical operation. **(Korean Circ J 2013;43:770-773)**

KEY WORDS: Sinus of Valsalva; Aneurysm; Ventricular outflow obstruction; Aortic valve insufficiency.

Introduction

Sinus of Valsalva aneurysm (SVA) is a rare anomaly with various clinical manifestations. Right SVAs are more common than left SVAs and if they rupture, they usually rupture into the right ventricle or right atrium.[1)] Occasionally, SVA protrusions can lead to hemodynamic obstruction of outflow tract on the underlying ventricle from the SVA origination.[2)3)] But non-traumatic rupture of right SVA complicated by left ventricular outflow tract (LVOT) obstruction has not yet been documented. We report a case of right SVA ruptured into the interventricular septum with resultant LVOT obstruction and aortic regurgitation in a previously healthy, middle-aged woman.

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Case

Fifty-four-year-old female visited outpatient clinic due to the worsening dyspnea experienced in the past month. Recently, she felt shortness of breath even during short distance walking. She also complained of intermittent coughing for the previous 6 months. She was diagnosed with essential hypertension 2 years ago, and has been taking single antihypertensive medication. She did not recall any other medical conditions which can explain her symptoms.

On her hospital visit, her blood pressure was 122/68 mm Hg, heart rate 95 beats per minute and respiration rate of 22 per minute. Physical examination was not remarkable except for loud to-and-fro murmurs which were audible along the left sternal border. Breathing sound was clear in both lung fields. The chest radiograph showed slight increases in cardiothoracic ratio without signs of pulmonary congestions. The electrocardiogram showed sinus rhythm with QRS configuration of right bundle branch block.

Initial laboratory studies revealed elevated levels of brain natriuretic peptide to 699 pg/mL, but the levels of cardiac markers including cardiac troponin and creatine kinase-MB isoenzyme were within normal limits. Routine blood cell counts and chemistry tests were also normal.

On transthoracic echocardiographic exam (TTE), basal part of interventricular septum was dissected by intrusion of a cystic cavity which communicated with coronary sinus of Valsalva. The cystic structure appeared in various sizes according to the cardiac cycle,

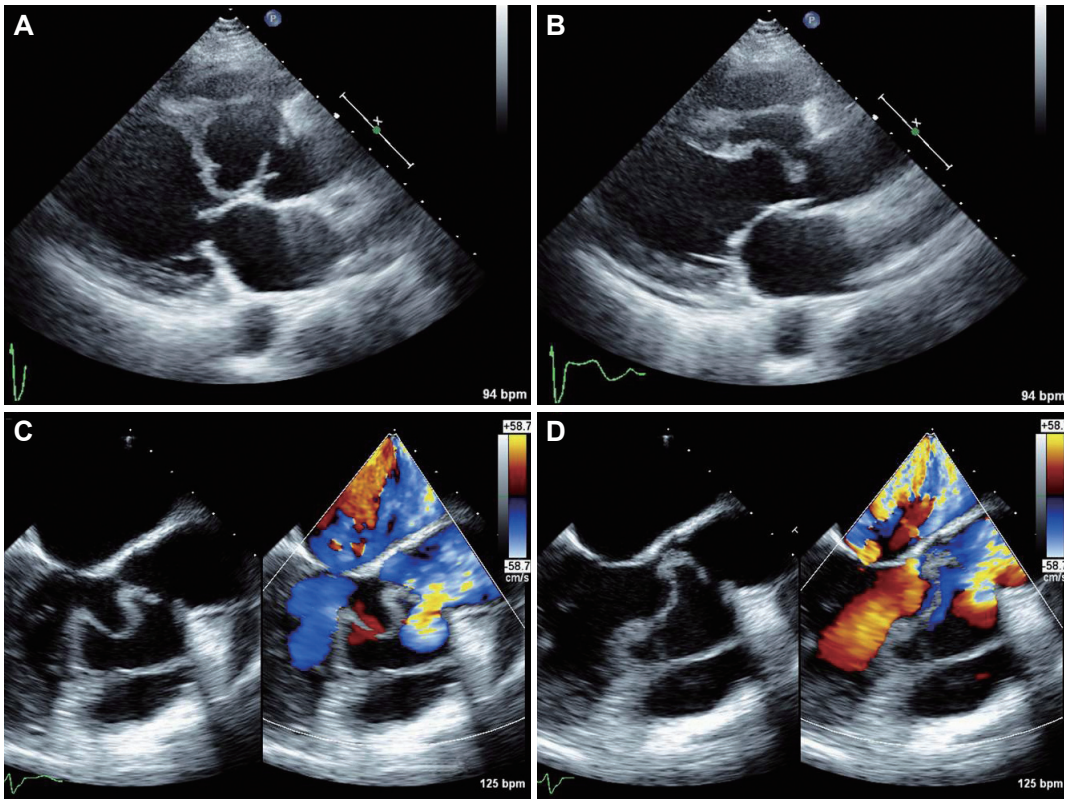

Fig. 1. Echocardiographic findings of SVA with interventricular dissection. Parasternal long axis view in TTE showing various sizes and appearances; increased size during diastole (A), decreased size during systole (B). Short axis view in TEE shows inflow from aortic root to SVA during diastole (C) and outflow during systole (D). SVA: sinus of Valsalva aneurysm, TTE: transthoracic echocardiography, TEE: transesophageal echocardiography.

increasing during diastole with protrusion into LVOT (Fig. 1A) and decreasing during systole (Fig. 1B). Transesophageal echocardiogram (TEE) also showed dynamic changes of the cystic structure with inflows (Fig. 1C) and outflows (Fig. 1D) between the aortic root and the cavity. However, no shunt flow was observed between left and right heart. Although the size of the protruding cavity decreased during systole, the hemodynamic obstruction of LVOT was recognized by Doppler echocardiography with peak pressure gradients of 55 mm Hg (Fig. 2). Additionally, aortic regurgitant flow of moderate degree was visible during diastole on both the TTE and TEE.

For further evaluations of the anatomical relationship between the SVA and surrounding structures, the cardiac multi-detector computed tomogram (MDCT) was performed. MDCT clearly demonstrated the main part of the SVA extending from the right coronary sinus (Fig. 3A). Interventricular septal dissection and cyst-shaped aneurysm throughout the cardiac cycle was best visualized through the scans (Fig. 3B and C).

Because of the disabling symptoms along with serious anatomic and hemodynamic deteriorations, clinical decision was made to offer prompt surgical corrections for the patient. Therefore, repair of interventricular septal dissection with concomitant aortic valve replacement was conducted. Right coronary cusp was also shortened

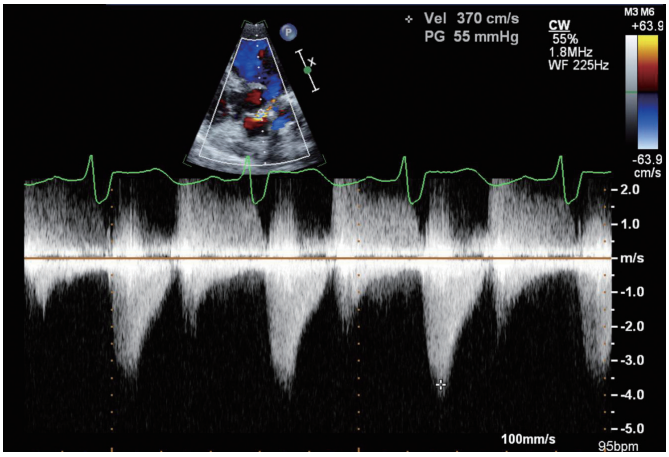

Fig. 2. Transthoracic echocardiography with continuous wave Doppler study revealed significant left ventricular outflow tract obstructions with pressure gradients of 55 mm Hg.

and diminished. Replacement of aortic valve through aortotomy was performed with 20 mm ATS mechanical aortic valve prosthesis (Medtronic Inc., Minneapolis, MN, USA). Histologic examination of the tissues revealed chronic degenerative changes (Fig. 4). There were no evidences of chronic or acute inflammations or infiltrative disorders. The patient was discharged uneventfully 9 days after sur-

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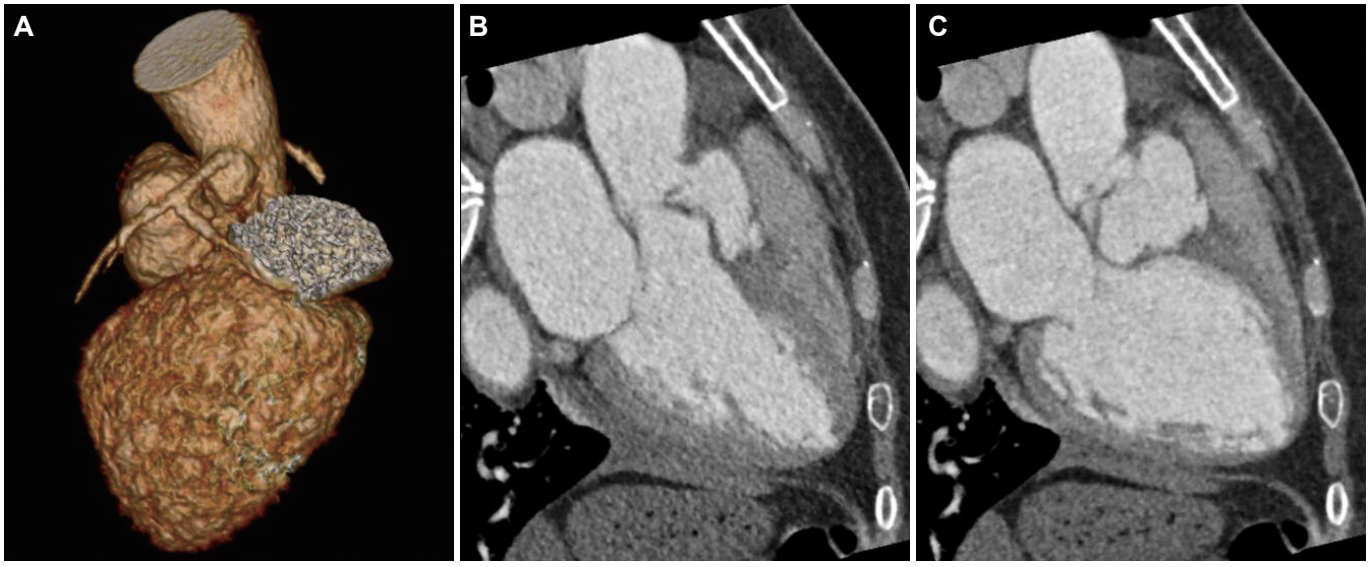

Fig. 3. MDCT findings. A: left lateral view of reconstructed image well visualizes the right SVA. B: MDCT shows interventricular dissection which forms cystic cavity at diastole. C: the interventricular cavity increases in size during diastole. MDCT: multi-detector computed tomography, SVA: sinus of Valsalva aneurysm.

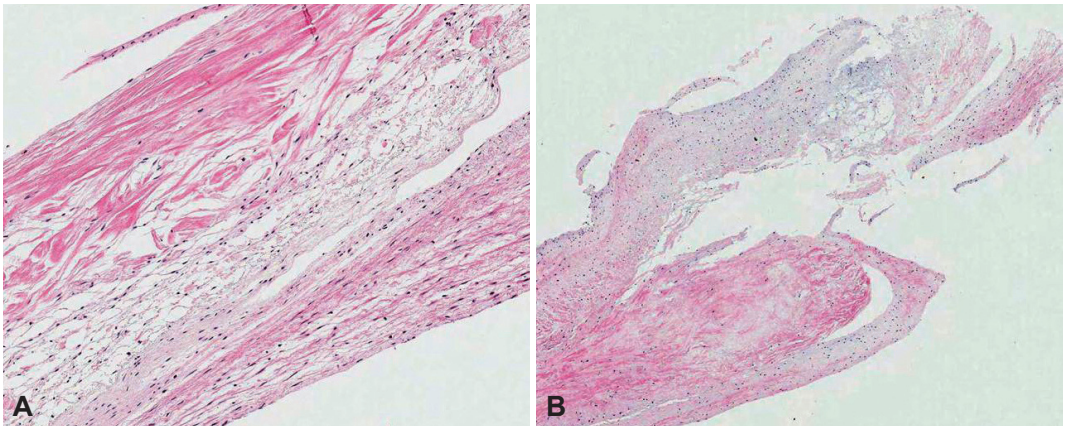

Fig. 4. Excised valvular tissues show dissected stroma by marked myxoid degeneration (A, H-E stain, ×40) and fibrillar degeneration with focal cystic changes in the central area of stroma (B, H-E stain, ×100).

gery, and did not have any complaints for 3 months thereafter.

Discussion

Sinus of Valsalva aneurysm is an uncommon anomaly of aorta, mostly derived from congenital defects of aorta media, and only rarely from acquired conditions.[4] The clinical manifestations of SVA vary widely, and they are related to ruptures or mass effects on adjacent cardiac structures.

The SVA in the present case originated from the right coronary sinus, which is the most prevalent site. When the SVA ruptures, they most commonly rupture into right ventricle, followed by right atrium, creating aortopulmonary shunt.[1)4)] In the present case, right SVA ruptured into interventricular septum by forming a cystic cavity. Rupture of SVA into interventricular septum is a rare complication, which comprises of less than 2% of SVA rupture.[1]

An interesting feature of this case is the hemodynamic LVOT obstruction caused by the protruding septal cavity originated from right SVA. The protrusion into LVOT generated significant pressure gradients of 55 mm Hg during systole, as measured by the Doppler echocardiogram. A case of unruptured SVA causing LVOT has been reported for the first time in 1998,[2] but it was a case of SVA originating from the left coronary cusp. Right SVAs dissecting the interventricular septum with protrusion into LVOT have been previously described.[5)6)] However, no hemodynamic measurements were made. SVA of the right origin with subsequent complications of the hemodynamic LVOT obstruction have not yet been reported to the best of our knowledge. The hemodynamic obstruction of LVOT in addition to acute volume overloading of the aneurysm rupture and aortic regurgitation might have aggravated the patient's symptoms.

Novel imaging techniques play an important role in the evaluation of SVAs. Conventionally, both TTE and TEE have been the first line diagnostic tool for anatomic and hemodynamic assessments of SVAs. Nonetheless, the cardiac MDCT and magnetic resonance imaging can provide additional information regarding 3-dimensional structures and anatomical relations between the SVA and adjacent organs.[7][8] In the present case, cardiac MDCT clearly demonstrated the configuration of interventricular septal dissection originating from right SVA and the anatomical relation with the aortic root and coronary arteries. Such data could provide valuable information for drawing up a therapeutic plan.

In conclusion, we report a case of right SVA ruptured into the interventricular septum which causes significant LVOT obstructions and aortic valve regurgitations at the same time, and was successfully managed through surgery. The MDCT and echocardiography made significant contributions for the evaluation of the case.

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