Three-dimensional echocardiography could distinguish a ventricular septal defect adjacent to asymptomatic ruptured sinus of Valsalva aneurysm

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KEYWORDS
Heart defects (congenital, ventricular septal defect); Aneurysms; Doppler ultrasound; Echocardiography (three-dimensional echocardiography)

Summary Sinus of Valsalva aneurysm (SVA) arises frequently in the right coronary sinus, and ventricular septal defect (VSD) is a prevalent coexistent cardiac abnormality. A 38-year-old asymptomatic male diagnosed with VSD on cardiac catheterization in his childhood, was referred to our hospital for the change in intensity of his cardiac murmur pointed out by his family physician. A grade V continuous murmur was auscultated with a thrill loudest at the forth left sternal border. Although, transthoracic and transesophageal echocardiography and cardiac catheterization have showed the ruptured right coronary sinus aneurysm, it was difficult to demonstrate coexistence of a doubly committed subarterial VSD. Three-dimensional echocardiography could provide clear images for diagnosis of the VSD closely adjacent to the ruptured SVA. The defect was confirmed at surgery.

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Introduction

SVA is caused by a congenital deficiency of fusion between the media of the aorta and the fibrotic tissue of the coronary sinus [1,2]. The disease often coexists other congenital abnormalities, such as VSD, bicuspid aortic valve, coarctation of aorta, etc. [3,4]. The 30–60% of the ruptured right coronary sinus aneurysm to the right ventricle accompanies a doubly committed subarterial VSD, which is relatively common in Asian patients [5–7]. Ruptured SVA is possible to be free from abrupt symptoms. The patients might tolerate sudden hemodynamic change when a congenital left-right shunt already coexists. Preoperative examination

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Figure 1  The ruptured SVA demonstrated by simultaneous recording of the 2D and color Doppler images on transesophageal echocardiography. (A) Long-axis planes and (B) short-axis planes. The right coronary sinus ruptured into the right ventricle. Although the large mosaic shunt flow appeared to be consisted of two different streams, the coexistence of VSD could not be visualized. LA: Left atrium, LV: left ventricle, RV: right ventricle, Ao: Aorta, RCC: right coronary cusp.

for the coexistence cardiac lesions should be carefully performed in patients with SVA. A concomitant VSD affects the surgical procedure which might need the additional aortic valve repair [8—10].

Three-dimensional echocardiography (3DE), which has advantages in spatial resolution, has become useful in the evaluation of various congenital heart diseases [11—13]. We experienced a case of a doubly committed subarterial VSD adjacent to a ruptured SVA, which could be clearly recognized by 3DE.

**Case report**

A 38-year-old asymptomatic male was referred to our hospital for increase in intensity of his cardiac murmur, which had been pointed out by his family physician in August 2006. He had been already auscultated a systolic heart murmur since his birth. He underwent right heart catheterization at the age of 10, and was diagnosed with VSD. However, surgical repair was not indicated because of a small left-right shunt on the oximetry run. The VSD was also demonstrated on echocardiogram when he was operated for pneumothrax at the age of 20.

On physical examination at admission, he was well developed with 170 cm in height and 65 kg in weight. Blood pressure and heart rate were 142/50 mmHg and 76 beats per minute, respectively. A grade V continuous murmur was auscultated with a thrill loudest at the forth left sternal border. A 12-lead electrocardiogram and a chest radiogram showed no obvious abnormalities. Transthoracic echocardiography with color Doppler study exhibited a large mosaic left-right shunt flow from the right coronary sinus to the right ventricle. On transesophageal echocardiography, a ruptured right coronary sinus was clearly demonstrated. Although, careful observations of long-axis planes suggested that the large mosaic shunt flow consisted from a fusion of two flows originated from the fractured right coronary sinus and the adjacent subaortic septal wall, separation of the overlapping jets was difficult (Fig. 1). In right cardiac catheterization, the mean pulmonary wedge pressure and the systolic pressure of the pulmonary artery were 10 and 29 mmHg, respectively. A cardiac output measured by Fick’s method was 3.45 l/min. On oximetry run, a significant step-up was confirmed in the right ventricle. The calculated $Q_p/Q_s$ was 2.59. Aortography showed the presence of a left to right shunt from the right coronary cusp to the right outflow tract, however, left ventriculography could not distinguish the coexistence shunt from the VSD.

We reconstructed the images of 3DE obtained from the commercially available machine (SONOS 7500, Phillips, USA) to recognize both the ruptured SVA and the doubly committed subarterial VSD. The accelerated color Doppler flow originating from the VSD could be clearly separated from the large shunt flow from the right coronary sinus (Fig. 2). Rendering of the VSD was performed just beneath the right coronary sinus at en face views (Fig. 3). The defect appeared to change its shape in each cardiac cycle.

In surgery, the right coronary sinus at the side of the left coronary cusp was ruptured to the right ventricle (Fig. 4A). The subaortic small size of VSD was confirmed underneath the hole of the right coronary sinus (Fig. 4B). Both defects were
Valsalva aneurysm rupture with VSD

Figure 2  Three-dimensional color Doppler echocardiography. (A) The shunt flow from the right coronary sinus into the right ventricle. (B) The acceleration flow originated from the VSD. The two different shunt flows were clearly visualized.

Figure 3  Sequential rendering (upper row) and color Doppler (lower row) images of the subaortic VSD in 3DE. Left, right, upper and lower side of the pictures is the direction of the left ventricular apex, the aortic cusps, chest wall and back, respectively. There was the VSD just beneath the right coronary cusp. The defect was closed in end-systole (c). Color-Doppler study demonstrated the shunt flow from the VSD during pre-systole (a) and systole (b). In end-diastole (d), the shunt flow disappeared, although the defect still opened. Solid arrow: VSD, dotted arrow: right coronary leaflet.

closed completely by patches with autologous pericardium.

Discussion

SVA occurs most frequently at the right coronary sinus, which ruptures usually into the right ventricle especially in Asian population. VSD is a common anomaly associated with SVA.

In recent development of echocardiographic technology, 3DE has been available for evaluating various heart diseases [14]. It provides the benefits for detecting congenital heart disease. In this case, the doubly committed subarterial VSD, which had been diagnosed in his childhood, could not
been visualized on transthoracic or transesophageal echocardiograms. It existed closely adjacent to the ruptured SVA. The shunt flow also overlapped the acceleration flow of the VSD. Reconstruction of the 3D on-line images made possible differentiating the two abnormal flows. En face views of the VSD could be rendered on the reconstructed 3D images. The orifice of this small VSD was dynamically opened and closed in each cardiac cycle. When closely coexistent structural abnormalities are present, 3DE display is excellent in spatial recognition.

As regards surgery, doubly committed subarterial VSD can cause overhang of SVA into the defect, which may partially obstruct the shunt flow [8,15]. However, the hemodynamic stress promotes rupture at the top of the aneurysm, and cause aortic regurgitation, for which aortic valve replacement is occasionally required even after patch closure [5—7,9,16]. The 3DE assessment may give great advantage to deciding operative procedures.

Finally, commercially available transthoracic 3DE leaves much to be desired in quality, frame rate and functional data to be measured. Nowadays, real-time transesophageal 3DE has been developed in clinical practice. It can provide more precise data for understanding the details in complex structural abnormalities.

In conclusion, 3DE is a useful modality for diagnosis of a VSD adjacent to a ruptured SVA in this case. The spatial recognitions were helpful for the surgical correction of the anomalies.

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