Incremental utility of Live/Real time three-dimensional transesophageal echocardiography in a case with ventricular septal aneurysm and hypertrophic obstructive cardiomyopathy: A case report

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Introduction

Ventricular septal aneurysm (VSA) is a very rare clinical condition with a reported frequency of 0.3% among patients with congenital heart disease (1). VSAs have a well defined association with ventricular septal defects (VSD). However, association with hypertrophic cardiomyopathy (HCM) is to be defined yet. We herein present a case with VSA and HCM, and use of Live/Real Time Three-Dimensional Transesophageal Echocardiography in differential diagnosis of this condition.

Case Report

A 40-year-old-woman was admitted to our clinic with exertional dyspnea lasting more than 3 months. Her past medical history and family history were unremarkable. She had a blood pressure of 115/80 mm Hg, heart rate of 110 beats/min, respiratory rate of 28/min, and body temperature of 37°C. Cardiovascular system examination revealed a harsh systolic murmur on the left sternal border. Resting ECG revealed sinus rhythm, pathologic Q waves in leads V1-V3, and nonspecific ST-T wave changes in the inferior leads. Laboratory investigations were within normal limits.

Transthoracic echocardiography (2-D-TTE) revealed left ventricular hypertrophy with interventricular septum (IVS) and posterior wall thickness of 15 mm and an ejection fraction of 65%. Systolic anterior motion of anterior mitral valve leaflet (SAM) was noted in the apical views (Fig. 1, Video 1). Continuous wave (CW) Doppler revealed a peak gradient of 133 mm Hg and a mean gradient of 73 mm Hg over LV outflow tract (LVOT) during resting conditions. Color Doppler examination revealed turbulence of blood flow below the aortic valve in an aneurysmal sac located in the membranous IVS (Fig. 2). Aortic valve was normal tricuspid valve and no other pathology which would cause secondary hypertrophy was detected. No direct connection between left and right ventricles was observed. In addition, severe mitral regurgitation (MR) secondary to SAM was seen. Right ventricular and atrial dimensions, right ventricular functions and pulmonary flow velocities were within normal range. According to these echocardiographic findings obstructive HCM, VSA with associated severe MR were the preliminary diagnoses.

Live/Real-Time Three Dimensional Transesophageal Echocardiography (RT-3-D TEE, Philips Medical Systems, Andover, MA, USA) was performed to clarify the anatomical properties of the VSA. The B-Mode 3-D datasets were cropped to show a sagittal cross section of the aneurysm (Fig. 3, Video 2). An aneurysm extending from the membranous septum in entrance of LVOT to junction of muscular and membranous septum was
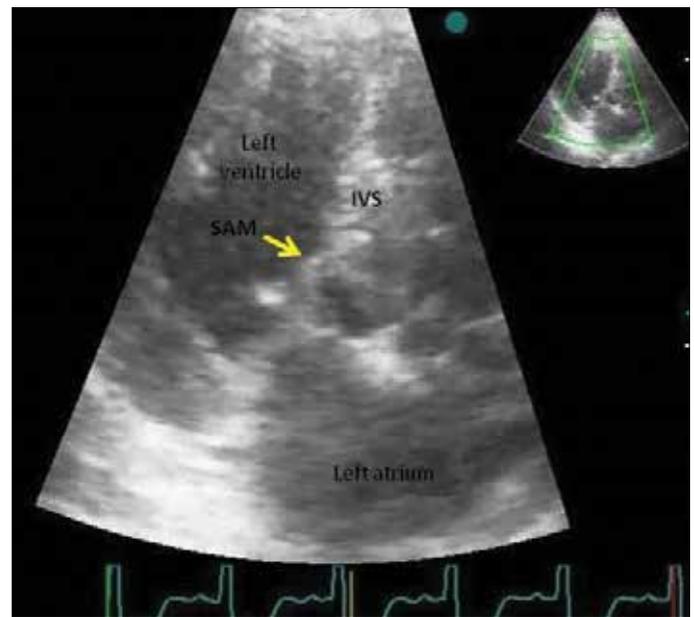

Figure 1. 2-D transthoracic apical 4-chamber view showing the close relation of mitral valve anterior leaflet with interventricular septum during systole compatible with systolic anterior motion (SAM)
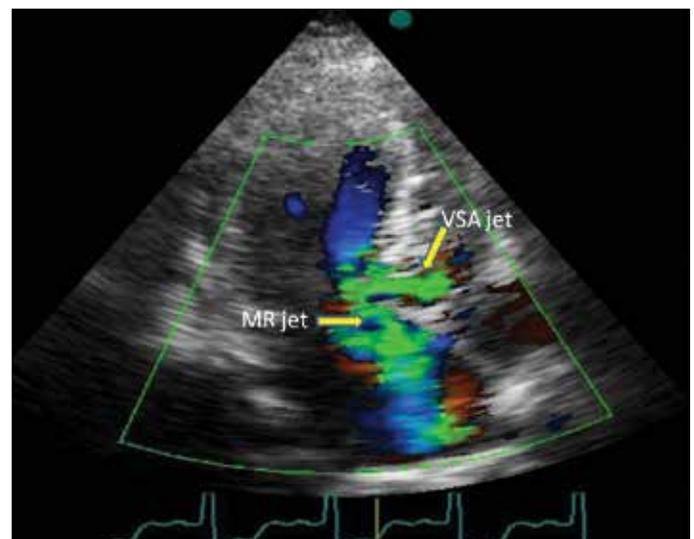

Figure 2. Color Doppler imaging showing the systolic turbulent flow of severe mitral regurgitation (MR) and of VSA

detected. 3-D-Advanced Quantification Plug-In (3-D-AdvQ) of the Q-LAB software was used to measure the volume of the aneurysm, which was 1.5 mL (Fig. 3). In 3-D color-flow imaging mode, it was clearly seen that there was no shunt through the aneurysm. The pathology was defined as VSA rather than diverticulum because of the wide neck and nearly hemispheric body with no myocardial tissue in the walls (2).

Patient underwent coronary angiography and left ventriculography in order to rule out coronary artery disease and confirm the diagnosis of HCM and severe MR. Coronary arteries were normal. In the right anterior oblique projection, severe MR was seen. Left anterior oblique projection with cranial angulation clearly showed the aneurysmal sac just beneath the aortic valve without a significant left to right shunt (Fig. 4, Video 3). During catheter pull-back, a maximal gradient of 136 mm Hg was observed between the aorta and LV cavity
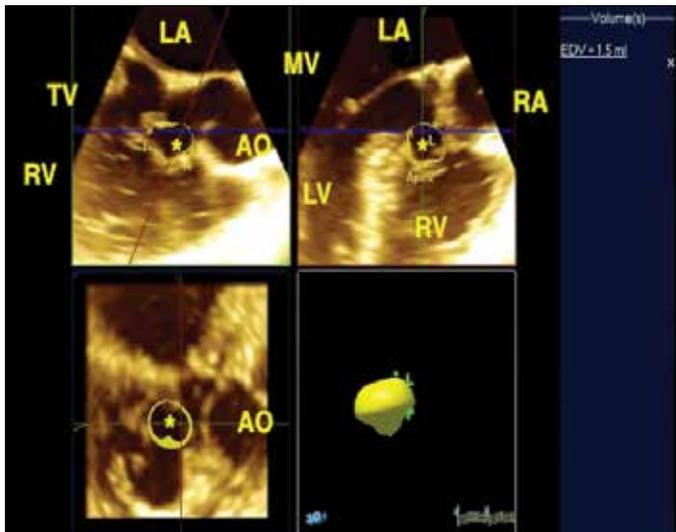
Figure 3. Cropped B-Mode 3-D dataset, clearly showing the anatomic relation of the aneurysm (asterisk) to AO - aorta; LA-left atrium; RV-right ventricle and TV-tricuspid valve and the volumetric measurement of the aneurysm (Asterisk) using 3-D-AdvQ plugin on Multiplanar Reformatting mode
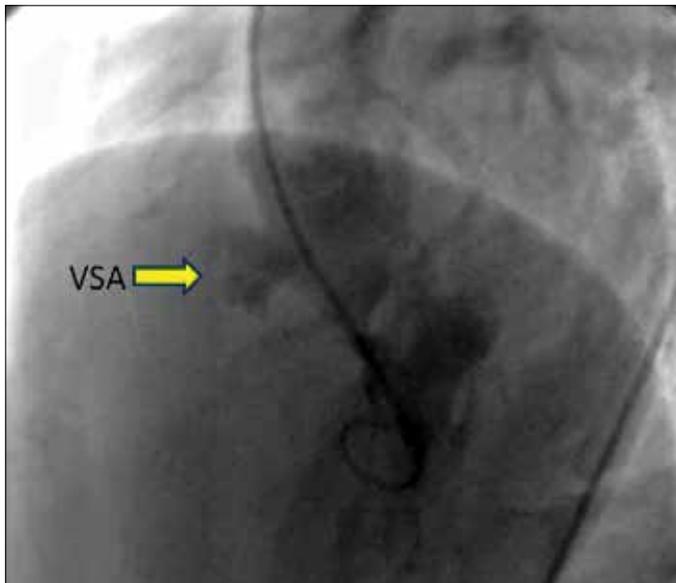

Figure 4. Ventriculography in left cranial projection showing the VSA located just beneath the aortic valve

(Fig. 5) and the diagnosis of hypertrophic obstructive cardiomyopathy was confirmed. As the patient had concomitant valvular disease and VSA, surgery was deemed more appropriate than septal ablation. Patient was referred for surgical treatment of severe mitral insufficiency, obstructive HCM and VSA, however the patient rejected the operation. The patient was discharged with acetylsalicylic acid, ß blocker therapy and was offered genetic counseling and family screening. The patient refused this offer. During one-year follow up she was in stable clinical condition.

Discussion

VSAs are very rare congenital anomalies with a variety of different clinical presentations and courses ranging from asymptomatic course
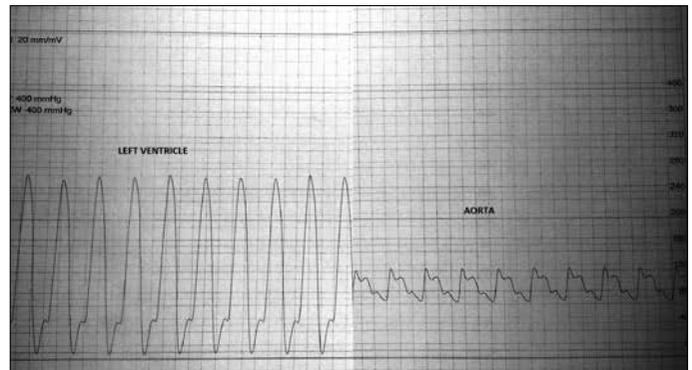

Figure 5. Catheter pullback pressure tracings showing the gradient difference between the left ventricle and the aorta

and incidental detection during work up for other causes, to AV block due to close proximity of the anatomical region to conduction system (3), arrhythmias (4) and sudden cardiac death, ventricular septal rupture, thromboembolic stroke or heart failure symptoms due to associated LVOT or RVOT obstruction.

Most common associated lesions with VSAs are VSD, aortic insufficiency, membranous subaortic stenosis, coarctation of aorta and AV canal defect, respectively (5). In literature, acquired cases of VSAs secondary to infective endocarditis and ischemic heart disease have also been reported (6, 7). According to our knowledge, concomitant diagnosis of VSA with HCMP has not been reported before.

Aneurysms usually range from 1 to 3 cm in diameter, are usually surrounded by fibrous tissue and may show apparent thick trabeculations when viewed from left ventricle (5). Kaplan et al. (8) demonstrated destruction of membranous tissue fibers and increased mucopolysaccharide content in aneurysmal wall.

Although many theories have been postulated regarding formation of aneurysms, the most accepted theory is closure of VSD with structures in close proximity to defect. In our patient, a prior diagnosis of VSD was not established during childhood. In addition, HCMP was not diagnosed before, neither. As both conditions are congenital in origin, the presence of a common developmental abnormality that would result in these two distinct structural abnormalities is theoretically possible. A genetic disorder involving these two conditions hasn't been defined yet. Alternatively, increased LVOT velocities with subsequent disturbance in flow dynamics might be the underlying mechanism, although differentiation between these two mechanisms cannot be made with current diagnostic modalities.

Following demonstration of gradient over LVOT, and turbulence in the area, 2-D TTE was insufficient in ruling out possible shunts, and in further clarification of dimensions and three dimensional anatomy of this condition. Chang et al. (9) reported the clinical utility of 3-D TEE in diagnosis of VSAs. Similarly, we clearly showed the boundaries of the aneurysm, ruled out a possible shunt through thin aneurysmal wall, and performed volumetric measurement of the aneurysm with RT-3-D TEE. RT-3-D TEE. Its more practical bed-side application, superior image quality, and quantitative analysis options, proved very useful in clarifying this case without need of further time consuming and expensive assessments such as computed tomography or magnetic resonance imaging (1, 10).

Conclusion

This is the first report in literature defining HCMP and VSA in the same patient. Utility of RT-3-D TEE in defining this condition added more

evidence to the substantial literature on usefulness of this assessment in diagnosis of complex anatomical pathologies of the heart.

Video 1. 2-D transthoracic apical 4-chamber view showing the systolic anterior motion of the mitral valve
Video 2. Three dimensional TEE image showing the anatomic relation of the ventricular septal aneurysm (Asterisk) to Aorta (AO), Left Atrium (LA), Right Ventricle (RV) and Tricuspid Valve (TV)
Video 3. Ventriculography in left cranial projection showing VSA located just beneath the aortic valve

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A rare cause of recurrent modified Blalock-Taussig shunt thrombosis: Antiphospholipid antibodies

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Introduction

The modified Blalock-Taussig (mBT) shunt is a palliative surgical treatment that increase pulmonary blood flow in patients with cyanotic congenital heart diseases. The incidence of mBT shunt thrombosis is reported to be among 1-17% (1). Several risk factors were defined about shunt thrombosis such as small age, low body weight, hypoplastic pulmonary arteries, small graft size and thrombophilia (1). We present a case of recurrent mBT shunt thrombosis associated with elevations in anticardiolipin antibody levels.

Case Report

A 22 month-old boy was admitted to our clinic with cyanosis. Arterial oxygen saturation (SpO_2) was measured 50%. A total blood count, partial thromboplastin time, prothrombin time, biochemical parameters were normal. Transthoracic echocardiography (ECHO) demonstrated that transposition of great arteries, L-malpozition of great arteries, ventricular septal defect (outlet), severe pulmonary stenosis (valvular, subvalvular). Systolic gradient between pulmonary artery and left ventricle was 90 mm Hg. Both right and left pulmonary artery was measured 6 mm. A right mBT shunt which using a polytetrafluoroethylene tube graft 5 mm in diameter was performed. After surgery we administered heparin infusion of 10 IU/kg/h and oral aspirin 5 mg/kg/day to prevent graft thrombosis. Several hours after surgery, his SpO_2 suddenly dropped below 50% and ECHO showed thrombotic occlusion of the shunt. He underwent angiography and first, heparin was given into the shunt, after then streptokinase and tissue plasminogen activator was administered. After shunt was successfully recanalyzed, immediately re-occluded and we performed balloon angioplasty promptly. Shunt was thrombosed repeatedly. Although stent placement into the graft, shunt flow remained insufficient. Therefore recanalization of the shunt with the same sized graft was performed surgically. Postoperatively shunt flow was inadequate and after heparin bolus was started, shunt flow increased dramatically. Because of this clinical picture, we suggest that thrombophilia and protein C, protein S, antithrombin III, prothrombin G20210A mutation, factor V Leiden mutation, folic acid, vitamin B12, homocysteine levels were measured. All of them were normal. In addition, we measured to antiphospholipid antibody (APA) levels and lupus anticoagulant and anticardiolipin antibodies were positive. We administered low-molecular-weight heparin and aspirin. He discharged with these therapy after five days and control laboratory tests showed that an undetectable level of APA. He was unremarkable during 13 months.

Discussion

Traditionally, diagnostic criteria of antiphospholipid syndrome (APS) in children was defined as: A combination of one of two clinical (thrombosis/recurrent abortions) and one of three laboratory features which are positive APA (lupus anticoagulant or anticardiolipin or anti-β2GP1) present on two occasions at minimum 12 weeks apart (2). However currently there is a quite controversy about diagnostic criteria of APS. Presence of anticardiolipin antibodies detect several other conditions, such as infants with atopic dermatitis, juvenile idiopathic arthritis, infections and vaccinations. It has been reported in healthy children, also (3). Furthermore some reports were published on seronegative patients who have the same clinical features with seropositive patients in APS (4, 5). Moreover there is a large variation about standardization and methods of measurement in APA levels. For these rea-