ECHOCARDIOGRAPHIC DIAGNOSIS OF SINUS VALSALVA ANEURYSM RUPTURE IN TWO PEDIATRIC PATIENTS*

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Sinus Valsalva aneurysm rupture (SVAR) is a rare cardiac abnormality that requires surgical correction when diagnosed. Previously, cardiac catheterization and angiography were thought to be necessary for its diagnosis. We present two pediatric cases of SVAR with subarterial ventricular septal defect (VSD) diagnosed noninvasively by echocardiography; surgical findings confirmed the diagnosis. In both of our cases the origin of SVAR was the right coronary sinus. The first case was ruptured into the right ventricular cavity; the second was ruptured into the right ventricular outflow tract.

Continuous murmurs heard during follow-up of children with VSD must alert the physician to this pathology. Combined two-dimensional, Doppler and color-Doppler echocardiography is an accurate, noninvasive method for diagnosis of SVAR. *Key words: sinus of Valsava aneurysm rupture, echocardiography, subarterial ventricular septal defect, children.*

Congenital aneurysms of the sinuses of Valsalva are thin-walled tubular sacs nearly always in the right sinus or the adjacent half of the noncoronary sinus[1]. Separation of the aortic wall media from the valve ring tissue causes this relatively rare lesion[2]. Rupture of sinus Valsalva aneurysm into right or, in rare cases, left heart chambers results in aortocardiac fistula[3]. Sinus Valsalva aneurysm rupture (SVAR) is rare in infants and children (0.1-3.5% of all congenital cardiac abnormalities)[4].

Although cardiac catheterization was considered necessary for accurate diagnosis of SVAR, recent reports indicate that SVAR can be diagnosed noninvasively by Doppler and color-Doppler echocardiography[5,6].

We herein present two cases of SVAR in the pediatric age group diagnosed echocardiographically and confirmed by surgery.

Case Reports

Case 1

A.K. was an asymptomatic 13-year-old boy with a known history of a small ventricular septal defect diagnosed by echocardiography in another center. On physical examination the left side of the precordium was prominent and

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hyperdynamic, and peripheral pulses were bounding. A loud continuous machinery murmur associated with a thrill was heard at the third and fourth intercostal spaces. Blood pressure was 110/70 mmHg. Electrocardiography showed normal for age sinus rhythm and QRS axis and left ventricular hypertrophy. Cardiothoracic ratio was 0.5 on chest x-ray and pulmonary vascular markings were normal.

Echocardiographic examination showed that all the heart chambers were slightly enlarged and ventricular contractions were normal (shortening fraction was 36%). A small ventricular septal defect on the subpulmonic region of the interventricular septum was present. The direction of the shunt through the ventricular septal defect was from left to right and a systolic gradient of 75 mmHg was measured between the two ventricles. Parasternal long and short axis and apical four chamber views demonstrated that the right coronary cusp of the aortic valve was prolapsed over the ventricular septal defect. A windsock-shaped tubular aneurysm arising from the right sinus of Valsalva and protruding into the right ventricle was detected (Figs. 1 and 2). Color flow mapping showed turbulent continuous flow originating from the aneurysm. Continuous wave Doppler interrogation of this flow revealed a continuous flow signal with a systolic peak of approximately 4 m/sec and a diastolic peak of 3.8 m/sec between aorta and right ventricle. Neither aortic regurgitation nor right ventricular outflow tract obstruction was found.

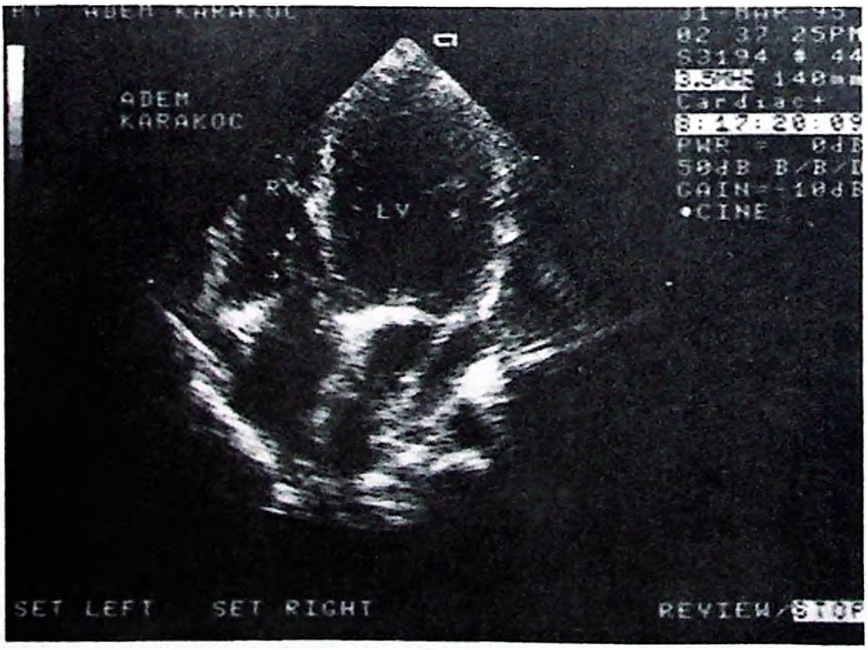

Fig. 1: Two-dimensional four chamber view of the first patient demonstrates the ruptured sinus Valsalva aneurysm protruding into the right ventricular cavity. LV: left ventricle, RV: right ventricle, Arrows: windsock-shaped aneurysm of sinus Valsalva protruding into the right ventricle.

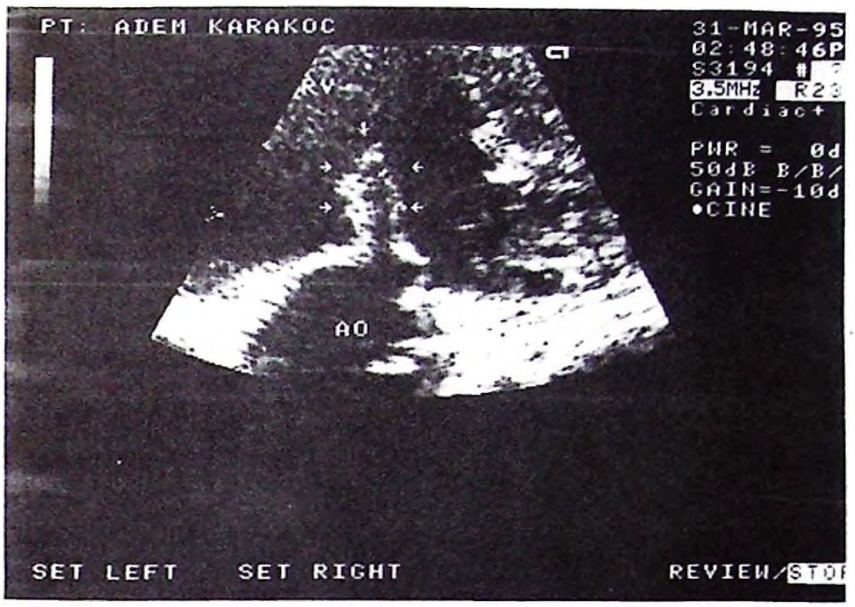

Fig. 2: Sinus Valsalva aneurysm protruding into the right ventricular cavity is seen in the parasternal short axis view of the first patient.

The ventricular septal defect was closed surgically with a dacron patch, and resection of the aneurysm pouch and primary repair of the rupture were performed. Echocardiographic findings were confirmed by surgery.

Case 2

A.T. was another asymptomatic eight-year-old boy without exercise intolerance. He was referred to our hospital for evaluation of a cardiac murmur heard during a routine examination. Physical examination revealed a hyperdynamic precordium and a 4/6th grade pansystolic murmur at the left sternal border associated with an early diastolic decrescendo type murmur; a thrill was palpable on the same region. Peripheral pulses were bounding. Cardiac silhouette was enlarged, pulmonary vascular markings were increased on chest x-ray and electrocardiography showed left ventricular hypertrophy.

Left atrium, left ventricle and aortic root were enlarged on echocardiographic examination. Contractility of the left ventricle was normal (shortening fraction 40%). Two dimensional echocardiography demonstrated a large subarterial ventricular septal defect. Left to right shunt was detected through the ventricular septal defect and the gradient between the left and right ventricles was 70 mmHg. A ruptured aneurysm of sinus Valsalva was protruding into the right ventricular outflow tract originating from the right coronary sinus of the aortic valve; a turbulent flow was detected in this region (Fig. 3). The leaflets of the aortic valve were deformed. Color Doppler echocardiography showed moderate aortic regurgitation. A mild obstruction with a 27 mmHg gradient at right ventricular outflow tract was present.

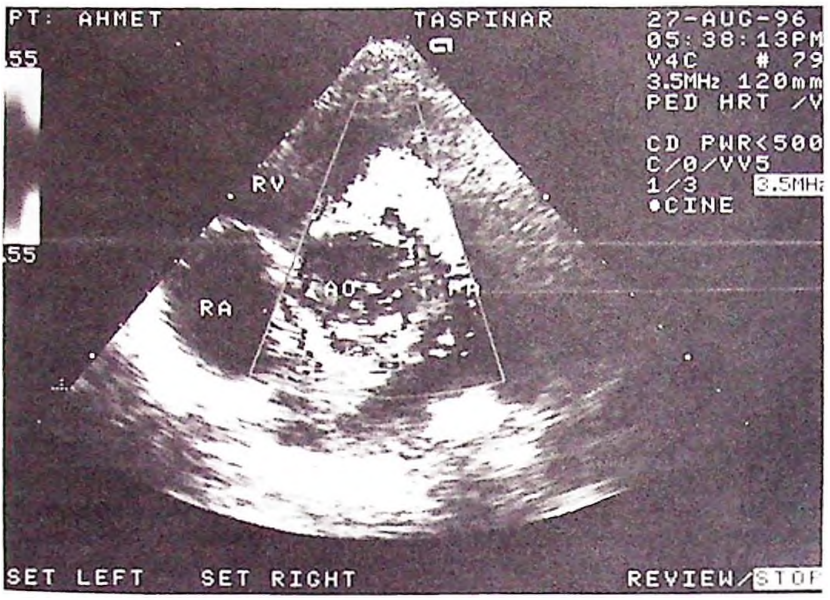

Fig. 3: Color flow imaging of the turbulent flow in the right ventricular outflow tract originating from the sinus Valsalva. Aneurysm is seen in the parasternal short axis view of the second patient. RA: right atrium, RV: right ventricle, Ao: aorta, PA: pulmonary artery.

At surgery, patch closure of the ventricular septal defect, repair of the SVAR and aortoplasty were performed. Anatomic features observed during surgery were consistent with the echocardiographic findings.

Discussion

Sinus Valsalva aneurysm rupture is a rare cardiac lesion. Its incidence among all patients who have undergone cardiopulmonary bypass is reported to range from 0.14-1.5%[7]. Though it has been reported in children as young as three years old, SVAR is more common in adults[1,2,8,9]. It is also more common in oriental populations in which subarterial ventricular septal defect is more prevalent[1]. Our patients are the fourth and fifth patients in our center to undergo open heart surgery with the same diagnosis in ten years (0.15% of all patients having open heart surgery). However, these are the first two patients to undergo surgery after echocardiographic examination without angiography.

Some of the patients may be asymptomatic, while in some, acute symptoms such as severe cardiac pain, dyspnea, tachycardia, exercise intolerance and congestive heart failure may occur[1]. It is thought that acute symptoms at the time of rupture may be less frequent when a subarterial VSD is also present[1]. There is little correlation between the size of the fistulous opening and the history of acute symptoms. Neither of our patients had acute symptoms but both had associated subarterial VSD which may explain the absence of symptoms.

Systemic diseases such as syphilis, infective endocarditis, Marfan's syndrome, cystic medial necrosis, atherosclerosis and trauma may cause SVAR[1, 7, 10-12]. No such disease was found in our patients.

Van Son et al.[1] found ventricular septal defect in 16 of their 37 patients with SVAR, and 15 of them were subarterial type ventricular septal defects. Both of our cases also had this type of ventricular septal defect.

Association of VSD with SVAR is reported in 30-50 percent of the cases in various studies[2, 4, 10]. Although presence of aortic valve prolapse associated with VSD and SVAR is reported rarely, it may be more common than indicates[3]. In both of our patients aortic valve prolapse was observed: aortic valve was competent in the first patient while moderate aortic regurgitation was observed in the second one.

Sinus Valsalva aneurysm rupture SVAR most frequently originates from the right coronary sinus (67-90%). Non-coronary sinus (8-25%) and left coronary sinus (0-8%) may be involved less frequently[2, 4, 10]. In both of our cases the origin of SVAR was the right coronary sinus. Aneurysms may rupture into the right ventricular outflow tract (60-85%), right ventricular cavity (20-25%), left ventricle (0-5.5%), or right atrium (9-66%)[2, 4, 10]. Our first case was an example of rupture into the right ventricular cavity and the second was ruptured into the right ventricular outflow tract. For SVAR in association with VSD, rupture into the right ventricular outflow tract is reported to be more common in childhood[2].

Sinus Valsalva aneurysm SVA may cause right ventricular outflow obstruction by mechanical compression. Aortic regurgitation is also detected in patients with VSD and aortic valve prolapse[13, 14]. Our second patient had both right ventricular outflow tract obstruction and aortic regurgitation.

Cardiac catheterization and angiography were considered to be necessary for diagnosis of SVAR in earlier reports[15]. Contrast echocardiography in combination with two-dimensional echocardiography was also used successfully in these patients[15]. Intracardiac ultrasonography is another method used for this purpose[16]. But with the development of Doppler and color-Doppler echocardiography and the improved resolution of two-dimensional echocardiography, presence of the aneurysm, location and size of the ventricular septal defect, origin of the aneurysm and cavity in which the rupture occurred can be demonstrated noninvasively[2, 5, 6].

In patients in which the sinus Valsalva aneurysm ruptures into the right ventricular cavity, the loud continuous signal in this area may cause difficulty for identification of right ventricular outflow tract obstruction. Similarly, small ventricular septal defects may not be detected because of this continuous flow. However, angiography may also miss some of these cases[2, 5, 6]. Color flow echocardiography is more valuable in detecting abnormal blood flow. In our cases we could define all anatomic features noninvasively by combining two-dimensional, Doppler and color-Doppler echocardiography, and our findings were confirmed by surgery.

In addition, our first case highlights the importance of follow-up in patients with small ventricular septal defects.

In conclusion, SVAR is a rare abnormality that requires surgical correction when diagnosed. Continuous murmurs heard during follow-up of children with VSD must alert the physician to this pathology and an echocardiographic examination must be performed. Combined two-dimensional, Doppler and color-Doppler echocardiography is an accurate, noninvasive method for diagnosis of SVAR.

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