Two-dimensional Echocardiographic Features of Atrial Septal Aneurysms

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SUMMARY  Aneurysms of the interatrial septum are rare. They have been associated with complications such as embolic phenomena and atrioventricular orifice obstruction. We describe two patients, one adult and one child, with atrial septal aneurysms that were diagnosed by real-time, two-dimensional echocardiography. Atrial septal aneurysms appeared as thin, localized outpouchings of the atrial septum that protruded into the right atrium and showed marked variations in their contour and size during the cardiac cycle. They could be differentiated from other intra-atrial structures such as tumor mass, large oesophageal valve and intra-atrial baffle by their relatively low reflectance, relationship to the atrial septum, considerable alterations in their outline during the cardiac cycle and characteristic patterns during peripheral venous contrast echocardiography.

ANEURYSMS of the interatrial septum are uncommon lesions that have been mistaken for intra-atrial tumors angiographically. Hence, a method is needed for making a definitive diagnosis of this condition. In this study we report two patients with atrial septal aneurysms diagnosed by real-time, two-dimensional echocardiography.

Case Reports

Case 1

Patient WL was a 68-year-old man in good health until 18 months before admission, when he noted the onset of shortness of breath while mowing his lawn. His exertional dyspnea progressively worsened and at the time of admission he could walk only 100 feet. Several weeks before admission he developed syncope while voiding. He had no other symptoms.

Physical findings included a blood pressure of 120/70 mm Hg and a regular pulse of 70 beats/min. No jugular venous distention was detected and the carotid upstroke was described as normal. The first heart sound was normal, S2 was widely split and an S4 was heard. A grade 3/6 systolic ejection murmur was audible at the base and radiated to the neck. A grade 2/6 diastolic decrescendo murmur was heard at the left sternal border. The lungs were clear. The clinical findings were consistent with the diagnosis of aortic stenosis and aortic regurgitation. A 12-lead ECG showed normal sinus rhythm, complete right bundle branch block and left ventricular hypertrophy with strain. Chest x-ray demonstrated marked cardiac enlargement and markedly increased pulmonary vascularity.

Echocardiographic Findings

M-mode echocardiography revealed a calcified aortic valve and a symmetrically hypertrophied left ventricle. The right ventricle was markedly enlarged (50 mm in end-diastole) and the interventricular septum demonstrated paradoxical anterior motion during systole. The mitral valve was normal. A band of abnormal linear echoes was also detected in the right atrium behind the prominent but structurally normal tricuspid valve and showed an undulating pattern in the cardiac cycle. Phasic differences in motion were present between the anterior and posterior limits of the echoes, and during atrial systole the anteriorly situated echoes moved toward the tricuspid orifice but did not prolapse into the right ventricular cavity (fig. 1).

Two-dimensional echocardiography was performed using a commercially available mechanical sector scanner (Picker) and confirmed the M-mode findings. In addition, parasternal and subcostal apical four-chamber views showed the presence of a thin, fragmentary linear echo in the right atrium whose convexity projected laterally to the right side. Considerable variation in the shape of the linear echo was noted during the cardiac cycle. This finding, as well as the absence of mass echoes in the right atrium, suggested the presence of a membrane-like structure rather than a solid mass. To further evaluate the nature of this lesion, an echo contrast study was performed by injecting 5 ml of 5% dextrose solution into a peripheral arm vein. Contrast echoes in the right atrium were limited by the linear echo, resulting in a filling defect (fig. 2) that showed marked variation in its contour in the cardiac cycle. It appeared to move relatively freely in the right atrium toward the region of the tricuspid valve, but did not prolapse into the right ventricle.

Cardiac catheterization revealed tight aortic stenosis, mild aortic regurgitation and evidence for an atrial septal defect. Because of the abnormal echocardiographic findings, a right atrial angiogram was performed and documented the presence of a right atrial filling defect that we initially thought represented a tumor. However, at operation no tumor was found. Instead, a thin, fenestrated, aneurysmally dilated in-
teratrial septum was found bulging into the right atrium (fig. 1). This was excised and the interatrial septal defect was closed with a Dacron patch. The aortic valve was replaced with a #16 Lillehei-Kaster valve. The patient was weaned from the pump successfully but developed intractable heart failure and died on the sixth postoperative day.

Case 2

JG was a 4-year-old white boy admitted to Strong Memorial Hospital for a Rastelli procedure and closure of a previously performed Waterston shunt (anastomosis of the ascending aorta to the right pulmonary artery). Cyanosis was noted when he was 2 days old.

Physical examination revealed an active child with a blood pressure of 90/40 mm Hg, pulse rate of 126 beats/min and regular, and respiratory rate of 32/min. He weighed 15.6 kg and was 96 cm tall. He was afebrile. Central cyanosis was noted. Neck veins were not distended. The apical impulse of the heart was diffusely palpable to the left of the sternum. The heart sounds were normal. A grade 3/6 systolic murmur was heard above the left sternal border. No gallop sounds or rubs were heard. All peripheral pulses were normal. A 12-lead ECG showed normal sinus rhythm and right ventricular hypertrophy. Cardiac catheterization demonstrated transposition of the great vessels, subpulmonic stenosis, ventricular septal defect and right-sided aortic arch.

Echocardiographic Findings

M-mode echocardiography revealed a prominent linear undulating echo behind the tricuspid valve. The great vessels were not visualized but both mitral and tricuspid valves appeared structurally normal. Real-time, two-dimensional echocardiography was performed with a wide-angle mechanical sector scanner (Advanced Technology Laboratories, Inc.). In the parasternal apical four-chamber view, a large segment of the atrial septum was seen protruding prominently into the left atrium during ventricular diastole as an undulating, curved linear echo with its convexity directed laterally. With the onset of ventricular systole, this segment of the atrial septum moved from the left atrium and bulged into the right atrial cavity, but the extent of the protrusion and the undulations were less prominent. Tricuspid and mitral valves appeared normal (fig. 3). Two transposed large vessels were identified. Multiple echoes were seen just below the posterior semilunar valve, consistent with the presence of an obstructing tissue mass in that region.

At the time of operation the findings observed during cardiac catheterization were confirmed. The pulmonary valve annulus was small, the pulmonary valve was bicuspid but not stenotic and there was dense, fibrous tissue obstructing the subpulmonic region. The Waterston anastomosis was closed. The ventricular septal defect was repaired so that the left ventricle ejected into the aorta, the connection between the left ventricle and the pulmonary artery was closed and a valved external conduit was placed between the right ventricle and pulmonary artery (Rastelli procedure). The postoperative course was complicated by cardiac tamponade that required surgical closure of a bleeding point in the right atrium on the tenth day. Subsequently, the patient developed
Figure 2. Aneurysm (AN) of the interatrial septum. The apical four-chamber view shows the atrial septal aneurysm as a thin, fragmentary, linear echo in the right atrial cavity. The basal portion of the atrial septum (AS) is intact. A peripheral i.v. injection of 5 ml of 5% dextrose in water produced contrast echoes in the right heart and outlined the atrial septal aneurysm as a filling defect in the right atrial cavity, which demonstrated marked changes in its outline during the cardiac cycle. CW = chest wall; RV = right ventricle; TA = tricuspid annulus; LA = left atrium; LV = left ventricle; VS = ventricular septum; MA = mitral annulus; I = inferior; S = superior; R = right; L = left.

Figure 3. Aneurysm of the interatrial septum. The apical four-chamber view shows prominent bulging of the atrial septum (AN) into the left atrium (LA) in ventricular diastole (upper panel). During ventricular systole (lower panel), the aneurysmal segment of the atrial septum bulged into the right atrial cavity (dotted). This patient had associated transposition of the great vessels, subpulmonic stenosis and ventricular septal defect. RV = right ventricle; TV = tricuspid valve; RA = right atrium; LV = left ventricle; MV = mitral valve; VS = ventricular septum; CW = chest wall; I = inferior; S = superior; R = right; L = left.
persistent hypotension, deteriorated progressively and died on the sixteenth postoperative day.

Postmortem examination confirmed the surgical findings. In addition, the patient had an atrial septal aneurysm in the region of the fossa ovalis, bicuspid aortic valve and a double-orifice right coronary artery.

Discussion

Aneurysms of the interatrial septum usually involve the region of the fossa ovalis. Often, they are asymptomatic and hemodynamically insignificant. However, serious sequelae have been noted that required surgical resection of this lesion. Stagnation of blood in large atrial septal aneurysms has resulted in clot formation, with subsequent cerebral and pulmonary embolic episodes. An aneurysmally dilated atrial septum may also protrude into the mitral or tricuspid orifice, resulting in obstruction simulating atroventricular valve stenosis. During angiocardiography, an atrial septal aneurysm may present as a filling defect in the right or left atrium that is indistinguishable from a tumor mass. Therefore, clinically, it would be valuable if a noninvasive technique such as echocardiography could be used to make a definitive diagnosis of this entity.

M-mode echocardiographic findings have not been found specific in the diagnosis of this entity. Generally, patients with atrial septal aneurysms demonstrate linear echoes in the right atrium behind the tricuspid valve (fig. 1). In an occasional patient in whom the aneurysm shows phasic bulging into the left atrium, linear echoes may be observed posterior to the aorta in the left atrial cavity. It is often difficult on the M-mode to differentiate an atrial septal aneurysm from an atrial tumor mass, although the echoes from an aneurysm are usually less prominent than those from a tumor and do not show significant protrusion into the valve orifice. Furthermore, a redundant and large eustachian valve may be mistaken for an atrial septal aneurysm on the M-mode.

Real-time, two-dimensional echocardiography appears to have the potential of making a definitive diagnosis of this entity. In our limited experience with this technique, atrial septal aneurysms present distinctive and characteristic features. They appear as localized, thin outpouchings of the atrial septum that regularly protrude into the right atrium because the left atrial pressure is normally higher than that in the right atrium during the cardiac cycle. In some patients the direction of the bulge is reversed in some phases of the cardiac cycle, particularly ventricular diastole, resulting in protrusion of the aneurysm into the left atrial cavity also. This finding appears to occur in patients with complex congenital heart diseases, such as transposition of the great vessels (our case 2) and tricuspid atresia and is presumably related to the phasic reversal of pressure relationship in the atria. The prominent variations in the shape and size of the thin, irregular echo outline produced by the aneurysm as it undulates in the atrium and its relatively low reflectance serve to differentiate it from an atrial tumor mass. Furthermore, the atrial septal aneurysm usually does not protrude significantly into the ventricle during diastole.

The eustachian valve may present as a prominent linear echo in the lower portion of the right atrium, but usually does not show prominent undulations when viewed from the apex. Also, when the atrial septum is studied in multiple planes, it would appear to be intact, with no evidence of outpouching. Peripheral venous contrast echocardiography may also help in the differentiation, because the atrial septal aneurysm (fig. 2), unlike the eustachian valve (fig. 4), will appear as a filling defect in the right atrium. An intra-atrial baffle inserted during the Mustard procedure for dextrotransposition of the great vessels may mimic an atrial septal aneurysm (fig. 5). However, a complete echocardiographic examination would demonstrate other segments of the baffle in other planes and dextrotransposed great vessels.

Aneurysmal bulging of the atrial septum is a well-known finding in patients with hypoplastic right-heart syndrome with tricuspid atresia (fig. 6) and can be suspected by M-mode echocardiography. In the only such case studied by real-time, two-dimensional echocardiography, the authors noticed abnormal echoes "in the position of the atrial septum" moving toward the right atrium during systole and toward the left atrium during diastole. However, their published illustration seems to demonstrate only a localized thickening of the atrial septum that does not show obvious bulging or bowing, and hence, no convincing evidence for the presence of an aneurysm. In our experience, localized thickening or abnormal echoes in the region of the atrial septum have been noted in patients with cardiomyopathy who have no evidence of associated atrial septal aneurysms.

Localized bulging of the atrial septum is indicative of an aneurysm formation because it points to localized weakening in the septal wall that results in the outpouching. Bulging of the entire atrial septum does not necessarily imply a totally aneurysmal atrial septal wall. Tei et al. described prominent, generalized bulging of the atrial septum into the right atrium in patients with atrial pressure overload, as in mitral regurgitation, and the septal bulge has been noted to disappear after mitral valve replacement, suggesting absence of aneurysmal involvement. Furthermore, to our knowledge, aneurysmal involvement of the entire atrial septum has not been documented pathologically.

Our patients had interesting associated cardiac lesions. Case 1 had calcific aortic stenosis and a left-to-right shunt through the fenestrated atrial septal aneurysm. Case 2 had transposition of the great vessels, ventricular septal defect, pulmonary valve stenosis, bicuspid aortic valve and a double-orifice
right coronary artery. In both cases, confirmation of the presence of atrial septal aneurysms and the associated cardiac lesions was obtained at surgery and/or at autopsy. Neither patient had tricuspid atresia or right-heart hypoplastic syndrome.

Our preliminary experience indicates that real-time, two-dimensional echocardiography is useful in identifying patients with atrial septal aneurysms.

Addendum

Since the submission of the manuscript, we have made prospective diagnosis of an atrial septal aneurysm in a patient with mitral stenosis using real-time, two-dimensional echocardiography. The echocardiographic features of the aneurysm were similar to those of case 1 and the findings were confirmed at surgery.
Figure 6. Atrial septum (AS) in tricuspid atresia. The subcostal four-chamber view demonstrates the atretic tricuspid valve (TV), hypoplastic right ventricle (RV) and generalized bulging of the entire atrial septum (AS) into the left atrium (LA). No localized aneurysmal protrusion is evident. The arrow points to a defect in the atrial septum produced by balloon septostomy in this patient. AW = abdominal wall; L = liver (hatched); RA = right atrium; MV = mitral valve; LV = left ventricle; VS = ventricular septum; R = right; L = left; I = inferior; S = superior.

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