Congenital Aneurysms of the Left Atrium: Recognition by Cross-sectional Echocardiography

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SUMMARY The two-dimensional echocardiographic features of three patients with congenital aneurysms of the left atrium are described. The aneurysm arose from the left atrial appendage in two patients and from the posterior left atrial wall in one. The aneurysms were characterized by their origin from an otherwise normal left atrium, a well-defined neck, their position within the pericardial space, and distortion of the left ventricular free wall by the aneurysmal body. The differentiation of these structures from other abnormalities of the left atrium are also discussed. Two-dimensional echocardiography is a safe and reliable method for diagnosing congenital aneurysm of the left atrium, and such studies should be considered in any patient with an otherwise unexplained abnormality on the chest radiograph.

CONGENITAL ANEURYSMS of the left atrium are rare but clinically important disorders. These aneurysms, which are considered to arise from focal areas of developmental weakness of the atrial wall,1 most frequently involve the atrial appendage,2-15 but may also arise from the body of the left atrium.2, 16-21 Although congenital, they do not usually become clinically apparent until the fourth decade or later, when complications such as cardiac arrhythmias and systemic embolization may occur.15 Because of these major complications, early diagnosis and surgical excision is mandatory.

An abnormal cardiac silhouette on the chest radiograph may suggest the anomaly, but is often mistaken for a cardiac tumor or pericardial cyst.19 Angiography is the established mode of diagnosis, the aneurysm is visualized by direct injection of contrast into the left atrium or by left-heart follow-through of contrast from a pulmonary arteriogram. In some cases, however, angiography has been misleading, and the correct diagnosis has been established only at thoracotomy or at postmortem examination.5, 11, 15-20

In this report, we describe cross-sectional echocardiographic findings in three patients with aneurysmal dilatation of the left atrial wall. In two patients, the diagnosis was confirmed at surgery, and in one the echocardiographic findings were definitive. Criteria for the diagnosis of this uncommon but potentially correctable condition are described.

Patients and Echocardiographic Findings

Patient 1

A 43-year-old woman was admitted to hospital with sudden onset of left hemiparesis, dysarthria and dysphasia. She had atrial fibrillation at a mean heart rate of 100 beats/min. There were no other abnormal physical findings, apart from the neurologic signs associated with her presenting symptoms. The chest radiograph
Serial presence of an evolving right ventricular regurgitation. The patient was given anticoagulant therapy; after her cerebral status improved, she underwent thoracotomy. A huge left atrial appendage was found in the pericardial space. The aneurysm was excised and contained no thrombus.

Patient 2

The second patient was a 39-year-old woman who had an abnormal cardiac silhouette on a routine chest radiograph. She was in normal sinus rhythm. Cardiac auscultation was unremarkable, as was the remainder of the physical examination. The radiographic abnormality was interpreted as a probable extracardiac cyst. Cross-sectional echocardiography revealed a large space to the left of the heart that markedly indented the left ventricular free wall and displaced the entire heart to a medial position. Echocardiograms in the apical four-chamber view showed that this space communicated with a normal-sized left atrium via a neck that was 2.5 cm in diameter (fig. 3A). The body of the structure was 6–7 cm at its largest diameter and extended laterally to the level of the cardiac apex. The mitral valve apparatus was normal. Scanning of the ultrasound plane from a left parasternal short-axis view of the aortic root toward the apex demonstrated an echo-producing mass that suggested thrombus in the aneurysm (fig. 3B).

Left ventriculography showed an extracardiac structure that appeared to compress the anterolateral wall of the ventricle. The left atrium was not opacified, as no mitral regurgitation was present. At thoracotomy, a giant aneurysm of the left atrial wall was found in the pericardial space. Adhesions to the surrounding mediastinal structures precluded resection of the aneurysm.

Patient 3

A 47-year-old woman who complained of palpitations was seen at a hospital clinic. Normal sinus rhythm was present. Cardiac auscultation demonstrated a mid-systolic click without murmur. The remainder of the examination was normal. A chest radiograph demonstrated moderate cardiomegaly. The ECG showed first-degree atrioventricular block and a QRS pattern of incomplete right bundle branch block. Cross-sectional echocardiography demonstrated a large cavity that lay in the pericardial space behind the heart and indented the posterior wall of the left ventricle. The cavity was best seen in the parasternal long-axis transducer view and communicated with a normal-sized left atrial cavity via a wide neck (fig. 4A). The body of the structure extended behind the heart toward the cardiac apex for 4 cm and was 2.5 cm at its largest diameter. The cavity was interpreted as a posterior left atrial wall aneurysm, and there were no echoes.

Figure 1. Plain chest radiograph from patient 1. The cardiac silhouette is abnormal, with a prominent convexity at the left basal aspect.

Figure 2. Cross-sectional echocardiogram in the apical four-chamber view from patient 1. The left atrial aneurysm (AN) is next to the lateral left ventricular wall and communicates with the left atrium (LA) via a neck (arrow). LV = left ventricle; RV = right ventricle; RA = right atrium.
suggestive of thrombus. The mitral valve apparatus was normal. This patient had an enlarged coronary sinus in the posterior atrioventricular groove that was clearly separate from the body of the aneurysm (fig. 4A). Contrast echocardiography after injection of indocyanine green dye into a left-sided arm vein confirmed the presence of a persistent left-sided superior vena cava, which drained completely via this coronary sinus to the right atrium. No contrast was seen in the left atrium or the cavity, which excluded the structure as part of the coronary sinus or a coronary sinus–left atrial connection.

The patient was advised to undergo cardiac rhythm monitoring and cardiac catheterization. However, the patient declined further evaluation and was lost to follow-up.

Discussion

Aneurysms of the left atrium may be congenital or acquired. They are considered to be congenital if they arise as a primary lesion from an otherwise normal atrial chamber and acquired if they develop in association with the massive left atrial dilatation that can occur with conditions such as mitral valve regurgitation or stenosis. Our three patients did not have significant mitral valve disease or left atrial enlargement; therefore, their left atrial aneurysms were considered isolated congenital anomalies.

The clinical findings in our three patients broadly conform to previous reports of this condition. The patients were women who became symptomatic in the middle decades of life. One patient had an unexpected radiographic finding, one had a history compatible
with atrial arrhythmia and one had a major systemic embolus. The anatomic location of the aneurysm was also consistent with previous reports. Fifteen cases of aneurysmal dilatation of the left atrial appendage and six cases of aneurysmal dilatation from the left atrial body have been described, a ratio approximately in accord with our experience.

Methods used to diagnose congenital left atrial wall aneurysms include plain chest radiography, M-mode echocardiography, radionuclide gated blood pool scanning and angiography. The radiographic appearance of an aneurysmal left atrial appendage may differ from that of an aneurysm of the atrial body. In all previously reported patients with aneurysms of the atrial appendage in whom it was obtained, the chest radiograph showed cardiomegaly or a prominent convexity at the left basal aspect of the cardiac silhouette. These findings are not specific, however, and may be misinterpreted. For example, the chest radiographs of patients 1 and 2 in our series suggested a left ventricular aneurysm and an extracardiac mass, respectively. Aneurysms arising from the body of the atrium may present radiologically as an anterior or posterior mediastinal mass. However, the cardiac silhouette may also demonstrate nonspecific cardiomegaly, as in patient 3.

M-mode echocardiography and radionuclide studies have been used in only one case to diagnose left atrial appendage aneurysm. The authors of that report drew attention to the nonspecificity of each technique. M-mode echocardiography showed the aneurysm as a space beside the heart that could not be distinguished from a pericardial or extracardiac cyst. Radionuclide gated blood pool scanning suggested an aneurysm of the pulmonary artery. Only by combining data from both studies could the correct diagnosis be deduced.

Angiography has been the most definitive method for establishing the diagnosis of congenital left atrial aneurysm. Angiographic definition of the abnormal anatomy depends upon the appearance of contrast material in the left atrium from the left-heart phase of a pulmonary arteriogram or after direct injection of contrast material into the left atrium. Since neither of these procedures is routine at cardiac catheterization, angiographic diagnosis implies that the condition was suspected before the procedure, which, from our experience and from previous reports, is unusual. Even if the left atrium does opacify with contrast material, thrombus may obliterate the aneurysm and defeat the angiographic demonstration of the abnormality. In addition, direct left atrial injection of contrast material may dislodge thrombus that might be present within the aneurysm.

Cross-sectional echocardiography is now a well-established technique for the noninvasive tomographic imaging of cardiac anatomy. However, its role in the diagnosis of congenital aneurysms of the left atrium and the characteristic echocardiographic features of this condition have not been reported. In each of our patients, the aneurysm was seen as a large cavity that lay next to the left ventricle and extended toward the apex within the pericardial space. The body of the aneurysm produced a clear indentation in the left ventricular wall in each case. In the two patients with aneurysms of the left atrial appendage, the aneurysm lay beside the anterolateral wall of the left ventricle and was best seen in the apical four-chamber view. This projection recorded the full extent of the structure and its point of origin from the lateral atrial wall near the atrioventricular groove. In patient 3, the aneurysm was behind the heart. In this position, its origin from the back wall of the left atrium and its extent was best seen in the parasternal long-axis view; the deformity in the posterior wall of the left ventricle that the aneurysm produced was most obvious in the short-axis view. The presence of intracavitary thrombus in patient 2 is in keeping with observations in patients at surgery or postmortem examination, and underlines the po-
tential danger of systemic embolus in all patients with this condition.

A finding of interest in each of these cases was the marked indentation of the left ventricular wall caused by the body of the atrial aneurysm. This feature has been recognized previously using angiography.\textsuperscript{4, 5} When measured directly, the pressure within the left atrium or aneurysmal body has not been elevated,\textsuperscript{6, 7} which means that the dilatation is not a consequence of raised intracavitary pressure. Therefore, this indentation of the left ventricle is presumably caused by expansion of the aneurysm into the relatively low-pressure pericardial space and subsequent distortion of the ventricle because of its presence within this confined area.

The main cross-sectional echocardiographic diagnoses from which true aneurysms of the left atrial wall must be differentiated are left atrial "aneurysms" of acquired origin, extrapericardial herniations of portions of the heart, solid or cystic paracardiac tumors and pericardial or extracardiac fluid collections, and an enlarged coronary sinus lying behind the heart.

A massively dilated left atrial appendage may be an acquired abnormality associated with the left atrial dilatation accompanying mitral valve disease, but can, of course, be distinguished from congenital aneurysms by the observation of an increased left atrial size or the structural mitral valve abnormality underlying the condition. Portions of the left atrial wall, pulmonary artery or left ventricle may occasionally herniate to an extrapericardial position in patients with partial defects of the left pericardium.\textsuperscript{26-28} However, this condition is distinct from true aneurysms of the left atrial wall, which are entirely intrapericardial in position.\textsuperscript{7} Although no reports of the cross-sectional echocardiographic findings of localized defects of the pericardium exist, the condition should be distinguished by its location outside the pericardial space and by the lack of ventricular distortion that intrapericardial compression might cause.

Loculated pericardial or pleural effusions and fluid-filled pericardial cysts should be distinguished by lack of communication with the left atrium. Similarly, a pericardial or paracardiac tumor such as lymphoma will not communicate with the left atrium. An enlarged coronary sinus receiving anomalous systemic or pulmonary venous return will sometimes appear echocardiographically as a space behind the heart.\textsuperscript{22, 23} In this instance, the space is confined to the posterior atrioventricular groove, and scanning of the ultrasound plane clearly shows that it enters the right atrium at the coronary sinus ostium.

In conclusion, cross-sectional echocardiography is a safe and reliable method for detecting congenital aneurysms of the left atrium. These aneurysms are characterized by their origin from an otherwise normal atrial chamber, a clearly defined communication with the atrial cavity and their interpericardial location with resultant distortion of the left ventricle. Although these aneurysms are rare, the associated complications and the relative ease of surgical resection suggest that such evaluation should be considered in any patient with an unexplained abnormality of the cardiac silhouette on the chest radiograph.

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