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ANSWER TO QUIZ – IMAGING ON PAGE 144 AND CASE DISCUSSION

Answer to quiz: A round cyst-like structure in the right atrium

Correct answer is #2: Atrial septal aneurysm (ASA) protruding from the left atrium into the right atrium.

This conclusion became clear after receiving other echocardiographic images, particularly the apical four-chamber view (Fig. 1). It is possible to notice on this image that in fact this formation is not a cyst, but a deep protrusion of the atrial septum in its central part from left to right. This protrusion remains practically the same by its size during both systole and diastole. Using the color Doppler we could not visualize the shunt of blood in the atrium from left to right side or vice versa, so it was not possible to prove the presence of a patent foramen ovale (PFO) or atrial septal defect, which often happens in such cases.

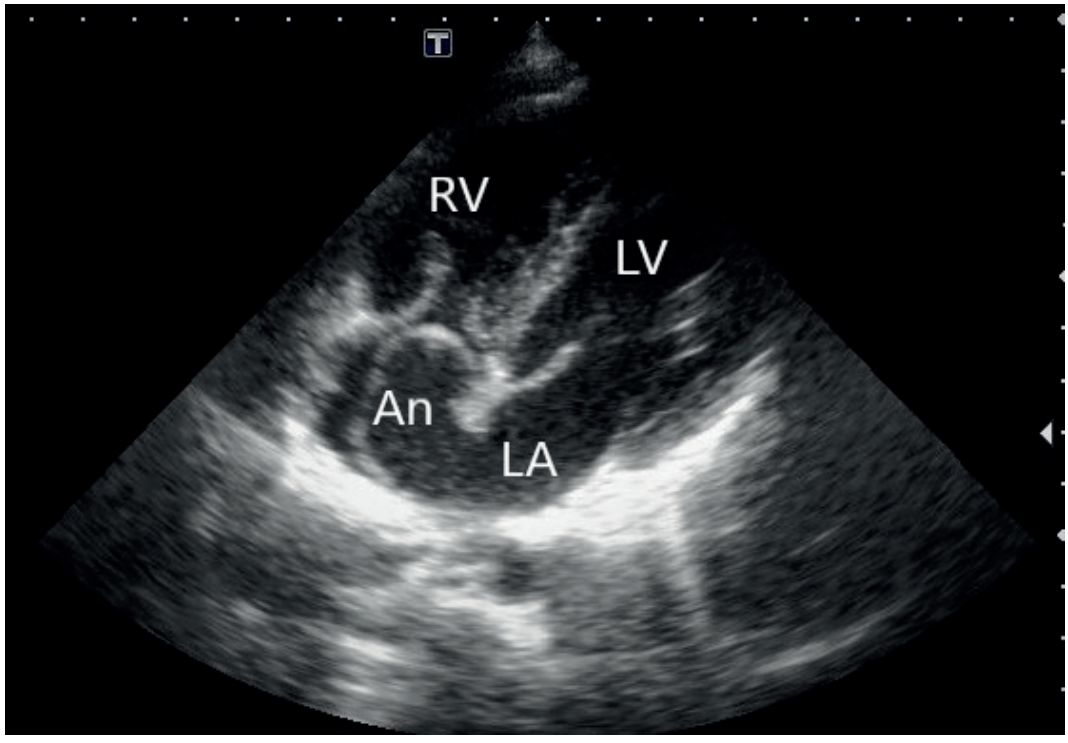

Figure 1. Apical four-chamber view. An – atrial septum aneurysm protruding from left to right atrium

This structure detected in the right atrium cannot be a blood cyst which has a different appearance on the echocardiogram. Cardiac blood cysts are primary cardiovascular benign tumors that are frequently found in the atrioventricular valves of newborn infants, but are extremely rare in adults. The origin of cardiac blood cysts is not understood and several hypotheses have been proposed. In our practice we dealt with two cases of blood cyst in adults, one of them is presented in Figure 2. They are not usually associated with other congenital heart pathology. Microscopically, cysts are a collection of blood cells surrounded by a thin membrane, which is lined with endothelium on the inside, and connective tissue on the outside. They are mostly single-chambered, although they can have membranes.

Address for Correspondence: Yuriy Ivaniv, Diagnostic Radiology Department, Danylo Halytsky Medical University, Lviv, Ukraine
Email: yivaniv@gmail.com
ORCID: 0000-0002-2153-9262
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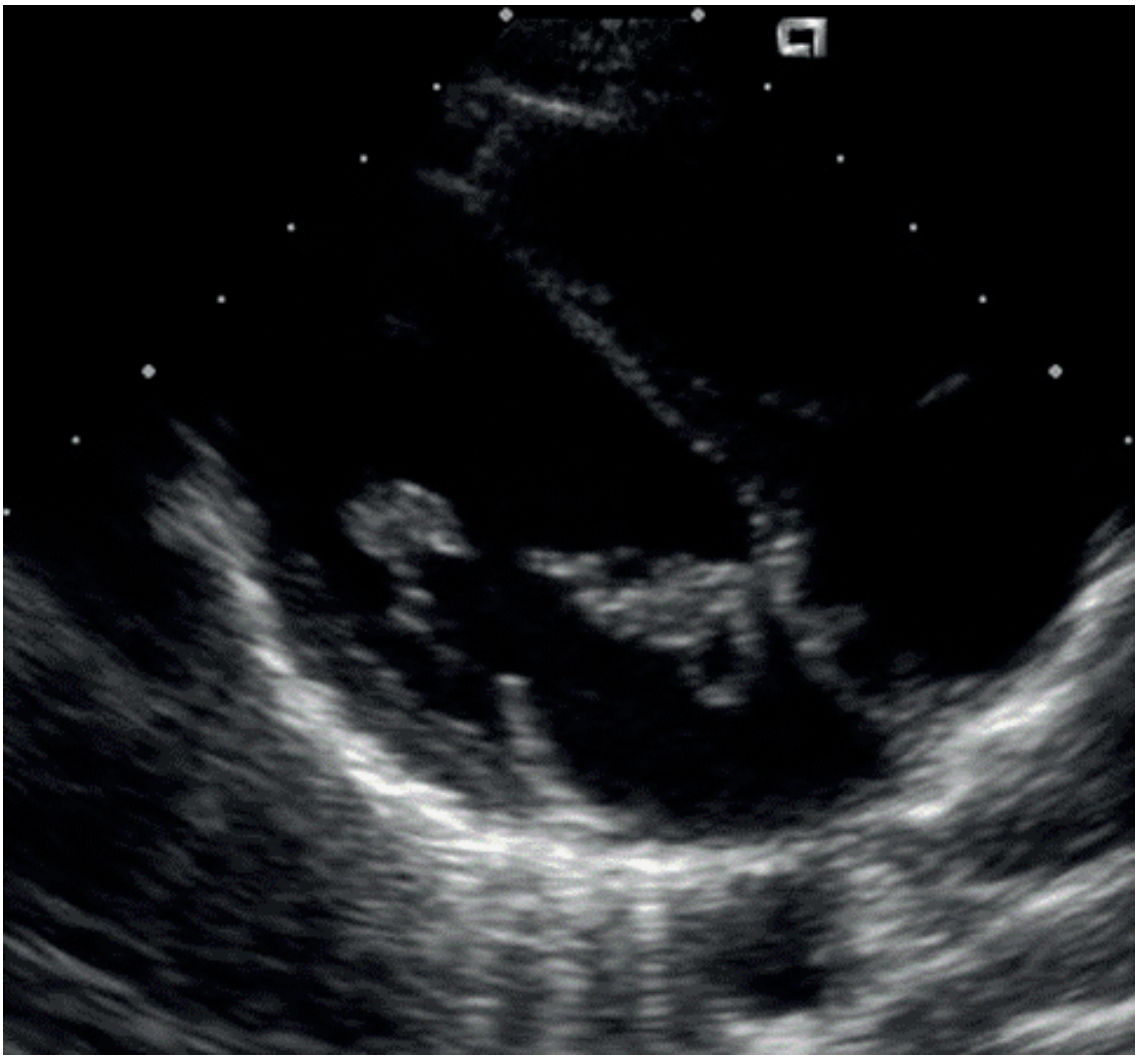

Figure 2. Blood cyst (indicated by arrow) attached to the anterior mitral leaflet at the atrial side

Echocardiography was first used to identify a blood cyst in 1983 and remains an excellent modality to diagnose this rare condition (1). Majority of reported cases of adult cardiac blood cysts in the literature were solitary and generally involved the valvular apparatus of mitral valve (2). The differential diagnosis of mitral valve cystic mass includes aneurysm or abscess as a complication of infective endocarditis, myxoma, cancer, hydatid or bronchogenic cyst. Although they are typically asymptomatic but rarely can cause complications.

As for echinococcal cysts they mostly present on echocardiographic imagine as a huge, well-defined intramyocardial multicystic mass with smooth contours and echo negative content (3). Any part of the heart may be affected but the most common location is the free wall of left ventricle or interventricular septum followed by atria and intracavity area (4). Diagnosis should be considered in patients coming from an endemic zone and presenting with an abnormal heart shadow on chest X-ray film. The hydatid cyst of the heart tends to grow compressing the neighboring cardiac cavities or structures. So it could cause displacement of the coronary vessels, rhythm disturbances and mechanical interference with heart valves and ventricular function.

Returning to the question of an atrial septal aneurysm, it should be noted that they are mostly focal, saccular outpouchings typically occurring they at the level of the fossa ovalis. These aneurysms can bow into one atrium only or may be bidirectional. They are found in 2% of healthy patients and 4% of patients undergoing transesophageal echocardiography. The diagnosis is made by imaging if protrusion of interatrial septum was more than 15 mm into the left or right atrium or phasic excursion more than 15 mm during the respiratory cycle, and the base of the aneurysm was at least 15 mm in diameter (5).

These aneurysms are typically congenital and can be associated with mitral valve prolapse, patent foramen ovale, and atrial septal defect. They are asymptomatic in themselves but may be complicated by atrial arrhythmias or thrombus formation and are often associated with intracardiac shunting.

Stroke is one of the most serious complications of ASA. Mattioli et al. (6) reported that ASA is the only potential cardiac source of embolism detected by transesophageal echocardiography in patients aged <45 years. It was also described a thrombus attached to the left side of an ASA that disappeared with anticoagulation therapy, but the aneurysm was eventually excised and repaired with an atrial patch because of the risk of recurrent thrombus and the need for lifelong anticoagulation therapy (7).

Various studies have demonstrated that the combination of ASA and PFO are associated with a higher risk of stroke. Cabanes et al. (8) reported that PFO and ASA are significantly associated with stroke in adults aged <55 years. But length of outpouching, extent of bulging, and incidence of spontaneous oscillations were similar in patients with and without previous cardiogenic embolism. However, associated atrial shunts were significantly more frequent in patients with possible embolism. This fact could indicate that paradoxical thromboembolism is the main mechanism of such complications. In some patients the cause of the interatrial shunt remains uncertain, but it might have been due to rupture of the weakened atrial septum as a result of a long-term bulging and stretching (9).

Atrial tachycardia is another frequent complication of ASA, with a prevalence of 18%-25% (10). The cause of arrhythmias in patients with ASA is not clear, but different cardiac pathology and abnormalities, such as atrial enlargement, arterial hypertension, left ventricular systolic or diastolic dysfunction and valvular prolapse might be responsible (11). It was established that atrial tachycardia in patients with ASA is not a risk factor for cardiac embolism per se.

The incidence of mitral valve prolapse associated with ASA is about 20% (12). Mitral valve prolapse and ASA might have a similar pathological basis, namely, a connective disorder involving fibrous cardiac tissue. It was supposed that connective tissue of an ostium primum atrial septum can become defective, especially when patients have myxomatous degeneration of the mitral valve. So a weakening of atrial septum might lead to the stretching and outpouching of it.

Treatment of atrial septal aneurysms is unclear although medical stroke prevention therapy is pursued in cases complicated by embolic stroke (5, 13).

Yuriy Ivaniv, Natalia Lozynska

Diagnostic Radiology Department, Danylo Halytsky Medical University, Lviv, Ukraine

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