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EDITORIAL COMMENT

Left Atrial Appendage Aneurysm

Knowledge and Gaps*

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Left atrial appendage (LAA) aneurysm (LAAA) is a rare condition characterized by a localized or diffuse dilatation of the LAA. Since it was first described by Dimond et al (1) in 1960, to date approximately 100 cases have been reported (2). The congenital form arises from dysplasia of the musculi pectinati and related atrial muscle bands. Acquired forms often occur in association with conditions leading to elevated left atrial pressure, including organic mitral valve diseases. LAAA has been seen in association with other congenital anomalies, such as atrial septal defect, ventricular septal defect, and Noonan syndrome (3).

Attending to actual literature we know a lot of things but have some doubts. The condition is seen in all ages, ranging from neonates to the ninth decade of life. Patients with LAAA may be asymptomatic, and the anomaly may be detected incidentally during cardiac imaging, or they may present with dyspnea, palpitations, and/or thromboembolic phenomena (2-4).

Transesophageal echocardiography (TEE) is superior to transthoracic echocardiography (TTE) for the detection of LAAA because TEE has a sensitivity of 90%. TEE should be performed in patients whose diagnosis is ambiguous during TTE examination, and TEE is also useful in the detection of intra-atrial or LAA thrombi. Cardiac computed tomography is the best choice for better definition of anatomy, to delineate the structure of the LAAA and its compressive effect on the surrounding structures (2).

Because LAAA predisposes patients to thromboembolism and atrial tachyarrhythmias, medical treatment with anticoagulation and antiarrhythmic drugs should be considered lifelong therapy. In view of this, the "consensus" is that surgical resection is the standard of treatment for LAAA because there have been no reported thromboembolic, arrhythmic, or other complications after aneurysmectomy. Anticoagulation therapy was discontinued in patients following surgical treatment of LAAA.

Given the rarity of this entity, we have several gaps of knowledge. First, there is no consensus definition of what constitutes an LAAA or what size is "aneurysmal." The size of the LAAA, as measured by TTE in the systematic review by Aryal et al (2), ranged between 2.2 × 1.1 cm and 13 × 10 cm. The smallest LAAA treated surgically was 2.9 × 2.2 cm. In a study of 500 normal autopsy hearts, Veinot et al. (5) reported the size of normal LAAs in different age groups and sexes. By using the +1 SD values in men as the largest normal dimension of the LAA, an LAAA could be defined as an LAA that has dimensions at the diameter of the orifice, width of the body, and the length of the LAA larger than 2.7, 4.8, and 6.75 cm, respectively (2,5). A standardized definition of LAAA is necessary for better reporting and management of this disorder.

Second, we need more information about the blood flow dynamics inside the LAAA and thromboembolic risk, to determine whether thrombogenicity depends on size, morphology, or concomitant atrial tachyarrhythmias. Aryal et al (2) published a logistic regression analysis, in 82 cases, to identify LAAA-related thrombus (left atrial or LAAA) formation and embolism. Variables included were age, sex, LAAA area in cm^2 (length × breadth), atrial fibrillation (AF) or flutter, and type of LAAA (congenital or acquired). By multivariate analysis AF or flutter was the only variable significantly associated with thrombus or embolism ($P < 0.05$). So, is a thromboembolic event

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possible in a patient with LAAA and sinus rhythm? Relevant evidence is provided by Pradella et al (6), in this issue of *JACC: Case Reports*. These investigators present the first known cardiac magnetic resonance (CMR) 4-dimensional (4D) flow analysis in a patient with a "giant" LAAA (5.7 × 5.3 × 8.2 cm) and sinus rhythm. Despite the confirmed absence of thrombus, the 4D flow sequence revealed significant flow between the left atrium and the LAAA (about 40 mL over 1 cardiac cycle) and LAAA "stasis maps" showed areas of high stasis, especially in the apex of the LAAA. These areas are at risk for thrombus formation, whereas conversely, the left atrium had no areas of high stasis.

Third, what is the potential risk of spontaneous rupture? We do not have information about that.

Fourth, although aneurysmectomy with extracorporeal circulation through median sternotomy, with pericardial reconstruction if necessary, is the widely accepted approach (4), some investigators have reported on the use of left lateral thoracotomy, without cardiopulmonary bypass, for resection, ligation, or stapling of the aneurysm (7). More recently, minimally invasive endoscopic aneurysmectomy with extracorporeal circulation has been performed (8).

The stasis maps provided by 4D flow CMR can be of great value as objective information in therapeutic decision making, whether medical or surgical, and to ascertain the need for permanent oral anticoagulation in patients with sinus rhythm and no thrombus or atrial arrhythmias but with a "large" LAA (no larger than 2.5 × 4.5 × 6.5 cm, to be more easily remembered) or LAAA (larger than 2.5 × 4.5 × 6.5 cm) without compressive symptoms and intermediate sizes.

In patients with a "large" LAA or a definitive LAAA, a history of AF or flutter or long paroxysmal atrial tachyarrhythmias, or documented thrombi in the left atrium or LAAA, oral anticoagulation is indicated. Moreover, if the patient has no contraindications, surgical treatment (aneurysmectomy) can be performed, to reduce the risk of arrhythmic and thromboembolic events and to enable the patient to discontinue lifelong anticoagulation therapy.

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