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Congenital Absence of Left Atrial Appendage in a Patient with Intracranial Hemorrhage

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Conflict of interest: None declared

Patient: Male, 78
Final Diagnosis: Congenital absence of left atrial appendage
Symptoms: None
Medication: —
Clinical Procedure: Transesophageal echocardiography • Cardiac CT
Specialty: Cardiology

Objective: Rare disease
Background: Intracranial hemorrhage is the most serious complication of anticoagulant therapy and is itself an absolute contraindication to further treatment.

Case Report: We present the case of a 78-year-old patient with permanent atrial fibrillation and previous intracranial hemorrhage during oral anticoagulation therapy, who was a candidate for percutaneous closure of the left atrial appendage. Transesophageal echocardiography and computed tomography showed absence of the left atrial appendage. The patient continued with single antiplatelet therapy.

Conclusions: Absence of the left atrial appendage is a very rare congenital condition usually found in patients scheduled for cardiovascular procedures and without clinical significance. The risk of thromboembolism is reasonably low but unknown.

MeSH Keywords: Atrial Appendage • Echocardiography • Heart Defects, Congenital • Stroke

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/894331>

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Background

Atrial fibrillation (AF) is the most common cardiac arrhythmia, and systemic embolization, particularly stroke, is the most frequent major complication. Anticoagulant therapy reduces the incidence of this complication, but overdose can cause intracranial hemorrhage (IH), which has an extremely high morbidity and mortality and is an absolute contraindication to further treatment with anticoagulant. In patients with AF and IH, closure of the left atrial appendage (LAA) reduces the risk of cardiac embolism at the most frequent site of thrombus formation.

Case Report

A 78-year-old male patient who was hypertensive, dyslipidemic, without previous cardiovascular surgery, and with permanent AF for the last 10 years, had IH during oral anticoagulation therapy with warfarin in 2012. Since then, he was treated with dual antiplatelet therapy with clopidogrel and acetylsalicylic acid and remained asymptomatic. He came to our attention for the pre-operative evaluation for LAA closure

because of his absolute contraindication to oral anticoagulant (CHA₂DS₂-VASC Score 5). We scheduled a transesophageal echocardiogram for morphologic evaluation of LAA and to exclude thrombus before the closure procedure. LAA was not visualized in ordinary and off-axis projections (Figure 1), raising the doubt of an aligned thrombosis occluding it versus an agenesis. No other cardiac anomalies were identified; right atrial appendage, descending aorta, and left ventricle were studied for other sources of embolism, without significant abnormality. Thanks to its optimal spatial resolution, a multidetector computed tomography scan was then performed, confirming congenital absence of the LAA (Figure 2). The patient continued with single antiplatelet therapy (clopidogrel).

Discussion

IH, primarily intracerebral and less frequently subdural or subarachnoid, is the most serious and lethal complication of anti-thrombotic therapy, causing approximately 90% of the deaths and most of the permanent disability in patients with warfarin-associated bleeding [1]. Approximately 60% of all strokes

Figure 1. (A–D) Trans-esophageal echocardiography; multiple views at mid-esophageal level were obtained not showing left atrial appendage. LA – indicates left atrium; LUPV – left lower pulmonary vein.

Figure 2. Multi-detector computed tomography imaging of the left atrium. (A) multiplanar reconstruction axial images of left atrium. (B) 3-dimensional volume-rendered images. LA – indicates left atrium; LAA – left atrial appendage; AscAo – ascending aorta; LV – left ventricle; PA – pulmonary artery; RUPV – right upper pulmonary vein; LLPV – left lower pulmonary vein.

in the National Institutes of Health stroke databank are caused by embolism [2], and cardiogenic embolism is responsible for 14–30% of ischemic strokes [3]. Among patients with nonvalvular AF, the vast majority of thrombi are located within or involve the LAA. The fibrillating LAA is the only area within the left atrium that is composed of pectinate muscle and can create an environment milieu conducive to blood stasis and thrombus formation. It is estimated that 90% of left atrial thrombi are located there [4] and percutaneous closure is therefore preferred in patients with absolute contraindication to oral anticoagulant. Two-dimensional transesophageal echocardiography (TEE) is the technique of choice to visualize LAA due to the proximity of the probe and it permits use of a higher frequency transducer, thus increasing spatial resolution. Three-dimensional TEE provides more specific information, which may be helpful in the differential diagnosis of agenesis with thrombus or other findings; it is particularly useful in the identification of muscular trabeculae. Absence of the LAA is a very rare congenital condition usually found incidentally in patients scheduled for cardiovascular procedures and without clinical significance. In our case this anatomic variation positively influenced clinical

decisions since it removes the main anatomic source of cardio-embolism. The risk of cardiac thromboembolism in this patient was reasonably low but unclear and the decision to leave the patient with single antiplatelet therapy, clopidogrel, is not based on evidence. We believe there are only 5 cases in the literature of LAA agenesis: they were patients scheduled for electrical cardioversion [5], for atrial fibrillation ablation [6–8], or for implantable cardioverter-defibrillator [6] and the discovery of this cardiac variation did not influence patient management or therapy decisions, as in our case.

Conclusions

LAA is the most common site of thrombus formation in patients with AF and closure of LAA is the preferred procedure in patients with previous IH caused by anticoagulant overdose. Congenital absence of the LAA is a very rare anomaly usually found incidentally and has no clinical impact on patient management. In our case it positively influenced choice of therapy and reduced thromboembolic risk.

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