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CASE REPORT

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Congenital absence of left atrial appendage in a patient with ischemic stroke and atrial fibrillation

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Abstract

Left atrial appendage (LAA) is a finger-like muscular extension of the left atrium, and it is the most prominent site for cardiac thrombus in patients with atrial fibrillation. Congenital absence of LAA could be incidentally detected in patient with ischemic stroke and atrial fibrillation. Although it is considered to be an extremely rare cardiac anomaly, its clinical significance remains unknown and there is no clear consensus in the management strategy in those patients. Therefore, we report a case of an incidentally noted congenital absence of LAA in a 68-year-old woman being planned for LAA closure.

KEYWORDS

atrial fibrillation, congenital absence, left atrial appendage

1 | INTRODUCTION

Left atrial appendage (LAA) is a small muscular extension of the left atrium (LA). It is closely related to the left pulmonary veins; anatomically, it lies anteriorly in the left atrioventricular sulcus in close proximity to the left circumflex artery.[1] LAA develops in third to fourth week of embryonic life from the left wall of the primary atrium and functions like a LA during the fetal life.[2] In adults, it is believed to function as a decompression chamber during elevated left atrial pressure including left ventricular systole or volume overload situations. It also contributes toward left atrial reservoir and contractile functions.[3] However, LAA is the most common source of thromboembolism in patients with atrial fibrillation (AF) and may be an arrhythmogenic source for the maintenance of AF.[4] According to the data analysis, there are 91% of intracardiac thrombosis associated with nonvalvular AF that are found in LAA.[5] Therefore, it is urgent to distinguish and prevent the occurrence of LAA thrombosis. Although the morphology of the LAA is highly variable among individuals, the absence of LAA is quite rare anatomical variation that clinical significance has not been discovered yet. We are going to report a case of congenital absence of LAA diagnosed by real-time 3D transesophageal echocardiography (RT-3D TEE).

2 | CASE REPORT

A 68-year-old woman with a medical history of hypertension, diabetes mellitus, cerebral infarction, and paroxysmal AF. Therefore, she was planned for LAA closure. Clinicians scheduled RT-3D TEE as a pre-operative procedure, for morphologic evaluation of LAA and to exclude thrombus before the closure procedure. LAA was not visualized in the whole process (Figure 1). In order to confirm, we checked patient's other relevant imaging history. She had no previous history of surgical or percutaneous LAA exclusion or occlusion, and computed tomography

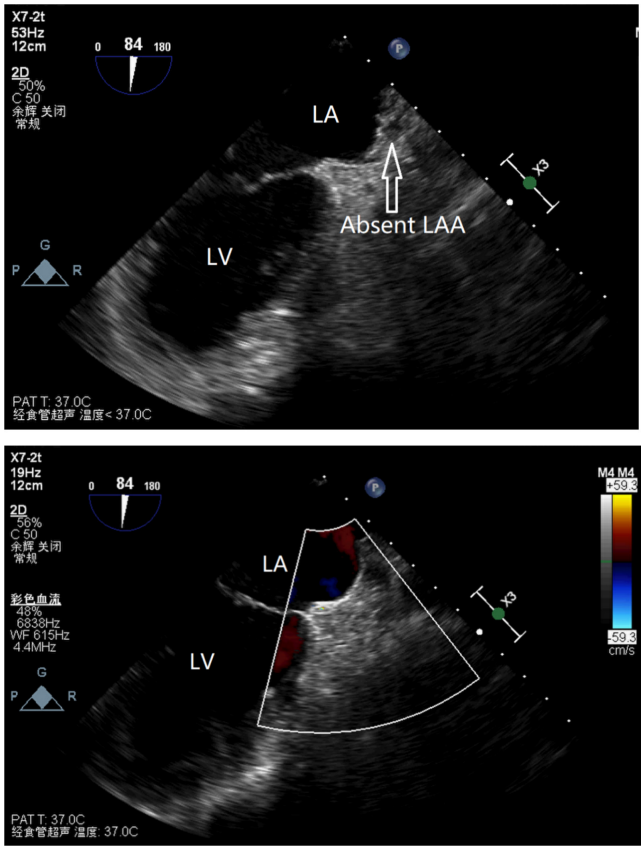

FIGURE 1 Missing LAA in the two-dimensional transesophageal echocardiography

showed no sign of imaging structure of LAA (Figure 2). Then, congenital absence of LAA was diagnosed and no thrombosis in LA was confirmed by RT-3D-TEE. Carotid plaques were found in a cursory scan of the carotid artery, and we assumed that unstable carotid plaques may be the main risk factor of cerebrovascular events in this case (Figure 3).

Anticoagulation therapy with warfarin was continued as per current guidelines since we have no data on anticoagulation management in congenital absence of LAA.

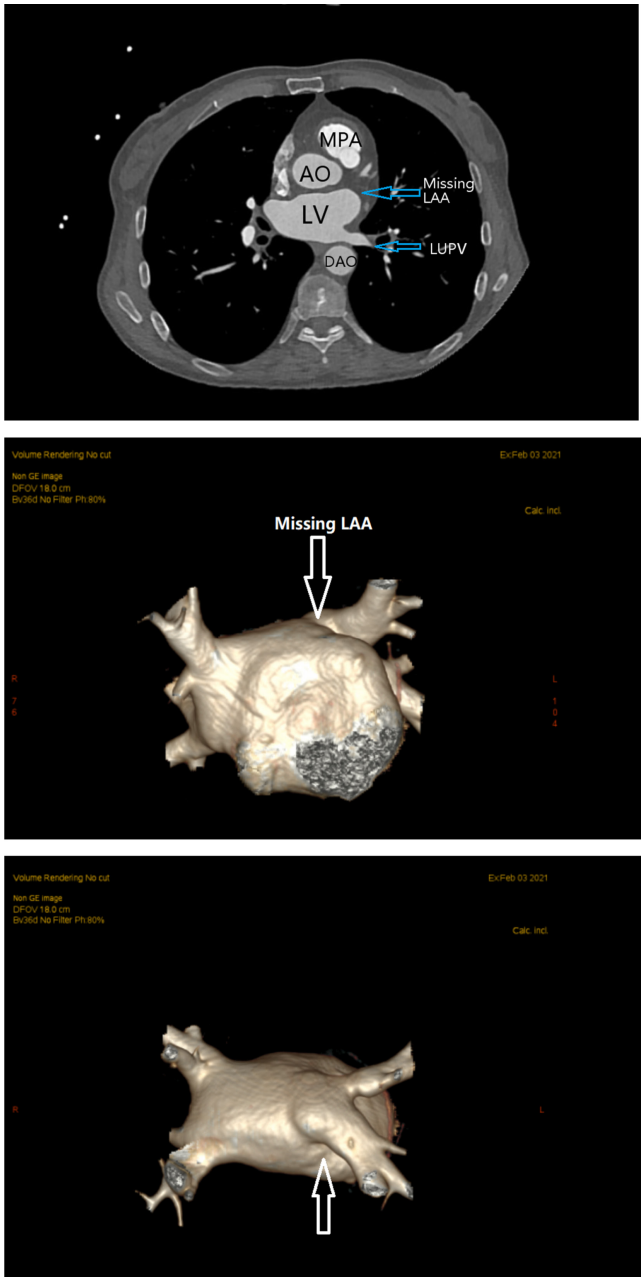

FIGURE 2 Computed tomography of pulmonary veins showed no left atrial appendage

3 | DISCUSSION

LAA is the only area within LA that is composed of pectinate muscle and creates an environment that is conducive to blood stasis and thrombus formation.[6] LAA also is a contractile reservoir and decompression chamber that acts as a suction during ventricular systole and as a conduit during diastole.[3] Functioning as an endocrine organ, when it stretched, LAA produces approximately 30% of atrial natriuretic peptides.[7]

The morphology of LAA varies greatly from individual to individual, which is generally divided into four types, including "chicken wing," "cauliflower," "cactus," and "windsock." Research showed LAA accounts for 91% of the thrombus sources in nonvalvular AF and 15%–38% in non-AF patients with a cardiomyopathy who have developed stroke.[5,8] A multicenter study had found that patients with the chicken wing morphology were significantly less likely to have an embolic event compared with those with cactus, windsock, and cauliflower morphologies.[9] In clinical practice, therefore, long-term oral anticoagulants are needed to prevent cerebrovascular accidents for those high-risk patients. Given that, AF-related thrombi occur predominantly in LAA, surgical and percutaneous procedures for LAA exclusion have been developed, especially

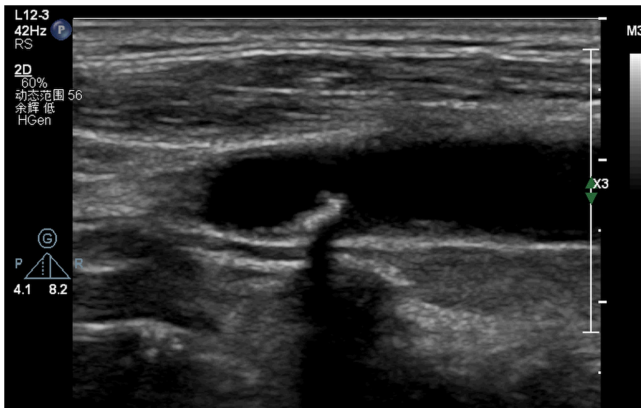

FIGURE 3 Plaque of carotid artery

for the patients who are not candidates for anticoagulation,[10] which include patients with prior hemorrhagic strokes and untreated bleeding disorders.

Congenital absence of LAA is quite rare cardiac anomaly, which can be found in multi-imaging processes intended for other purposes. The accurate prevalence and incidence of this condition is unknown. Its diagnosis needs to be considered on detail evaluation of patient's surgical and medical history as total thrombotic occlusion, uncommon anatomical features, surgical or percutaneous exclusion, and poor imaging quality could cause misdiagnosis. Additionally, considering the variations of LAA position and morphology, multimodality imaging is often needed for confirmation.[11] TEE is the technique of choice to visualize LAA due to its higher spatial resolution and real-time performance. 2D TEE can more accurately evaluate the morphology and function of LAA, as well as the adjacent structure of LAA; meanwhile, 3D TEE provides more specific information, which may be helpful in the differential diagnosis of LAA with thrombus or other findings. RT-3D TEE is a novel and valuable imaging modality in the percutaneous catheter-based LAA occlusion in AF patients, which could be recommended for routine clinical application.[12]

Theoretically, the risk of embolic events in patients with AF with congenital absence of LAA is low, but clinical significance of the anomaly was not elucidated. In our case, it is worth mentioning that various etiologies can evoke stroke events and defining stroke mechanisms is crucial for effective stroke prevention.

AUTHOR CONTRIBUTIONS

Mayire·aobuli was mainly responsible for article writing and image processing. Li Li Jia involved in image processing. Nuliya·Yasen contributed to collecting patient data and dealing with the submission process. Gui-ming Zhou contributed to technical support. Xin Du contributed to technical and data support.

CONFLICT OF INTEREST

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

CONSENT

I confirm that written patient consent has been signed and collected in accordance with the journal's patient consent policy, and that I have added a patient consent statement asserting this at the bottom of the manuscript's title page. I will retain the original written consent form and provide it to the publisher if requested.

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