Congenital Absence of Left Atrial Appendage in a Patient with Ischemic Stroke

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Abstract

We report a case of congenital absence of left atrial appendage in a 68 years old female who was going to proceed a left atrial appendage occlusion for ischemic stroke in whom we observed the absence of left atrial appendage by Real-Time 3D Transesophageal echocardiography (RT 3D TEE).

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Abstract :

The left atrial appendage (ALL) is a fingerlike muscular extension of the left atrium and its congenital absence is considered as an extremely rare circumstance which can be found accidentally in the imaging processes intended for other purposes .Therefore we report a case of congenital absence of left atrial appendage in a 68 years old female who was going to proceed a left atrial appendage occlusion for ischemic stroke in whom we observed the absence of left atrial appendage by Real-Time 3D Transesophageal echocardiography (RT 3D TEE).

Key words: Left atrial appendage;Transesophageal echocardiography

Introduction :

The left atrial appendage (ALL) 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]. Left atrial appendage develops in 3rd -4th week of embryonic life from the left wall of the primary atrium and functions like a left atrium 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 towards left atrial reservoir and contractile functions [3]. The LAA, however, 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 the LAA[5].Therefore it is urgent to distinguish and prevent the occurrence of left atrial appendage thrombosis. Although The morphology of the left atrial appendage is highly variable among individuals, the absence of ALL 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).

Case report :

A 68-year-old woman with a medical history of hypertension (HTN) for 2 years, diabetes mellitus (DM) for 15 years, cerebral infarction for 5 years, and paroxysmal atrial fibrillation for the past 13 years, additionally there were radio frequency catheter ablation for three times for rhythm control in the past 10 years. Because of frequent palpitations, the left atrial appendage was planned to be occlusion, clinicians scheduled a transesophageal echocardiogram, 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 checkup patient‘s other relevant imaging history, the patient had no previous history of surgical or percutaneous left atrial appendage exclusion or occlusion, and there was Computed tomography angiogram of the pulmonary veins proceeded before which showed no sign of imaging structure of LAA (figure 2). Then the congenital absence of left atrial appendage was diagnosed and no thrombosis in the left atrium was confirmed by RT 3D TEE. Carotid plaques were found in a cursory scan of the carotid artery before transesophageal ultrasound, 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 left atrial appendage.

Discussion:

The LAA is the only area within the left atrium 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[7]. Functioning as an endocrine organ, when it stretched, the LAA produces approximately 30% of atrial natriuretic peptides [8]

The morphology of LAA varies greatly from individual to individual which is generally divided into 4 types, including "chicken wing," "cauli-flower," "cactus," and "windsock". Research shows the LAA accounts for 91% of the thrombus sources in nonvalvular AF and 15% to 38% in non-AF patients with a cardiomyopathy who have developed stroke [9,10]. A multicenter study has found that patients with the chicken wing morphology are significantly less likely to have an embolic event compared to those with cactus, windsock, and cauli-flower morphologies [11]. In clinical practice, therefore, long term oral anticoagulants are needed to prevent cerebrovascular accidents for those high risk patients. Given that AF-related thrombi occurs predominantly in the LAA, surgical and percutaneous procedures for LAA exclusion have been developed especially for the patients who are not candidates for anticoagulation[12], which include the patients with prior hemorrhagic strokes and untreated bleeding disorders.

The 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, as well as poor imaging quality could cause misdiagnosis. Additionally, considering the variations of LAA position and morphology, multimodality imaging is often needed for confirmation [13]. TEE is the technique of choice to visualize LAA due to its higher spatial resolution and real-time performance. Two-dimensional TEE can more accurately evaluate the morphology and function of the left atrial appendage, as well as the adjacent structure of the LAA, meanwhile Three-dimensional 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 [14].

Theoretically, the risk of embolic events in patients with AF with congenital absence of left atrial appendage is low, but clinical significance of the anomaly was not elucidated. In our case, It's worth to mention that various etiologies can evoke stroke events and defining stroke mechanisms is crucial for effective stroke prevention.

Author Contributions:

1. mayire·aobuli : Mainly responsible for article writing and image processing
2. Li Li Jia:image processing
3. Nuliya*Yasen:Collecting patient data and dealing with Submission process
4. Gui-ming Zhou: Technical support
5. Xin Du: Technical and data support

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Figure legends:

Figure 1: missing LAA in the Two dimensional transesophageal echocardiography

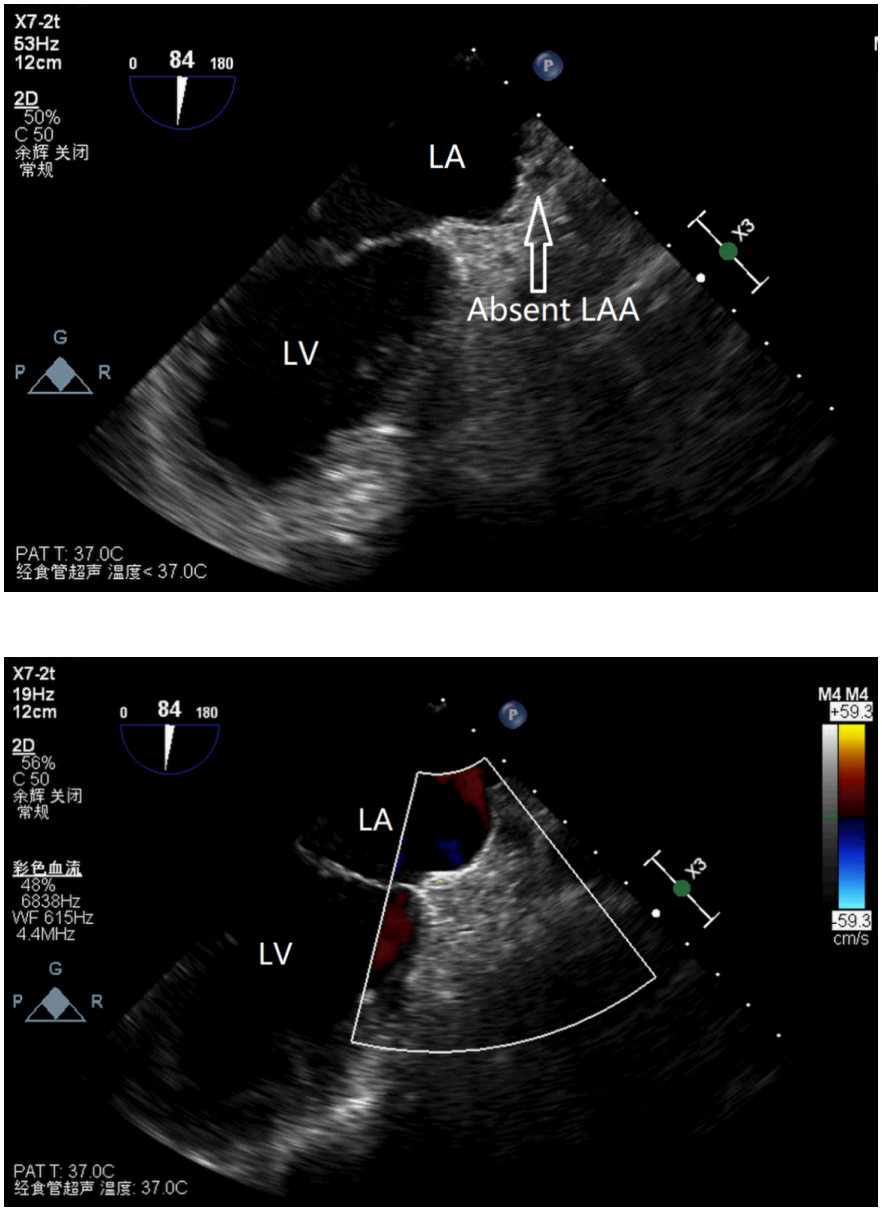
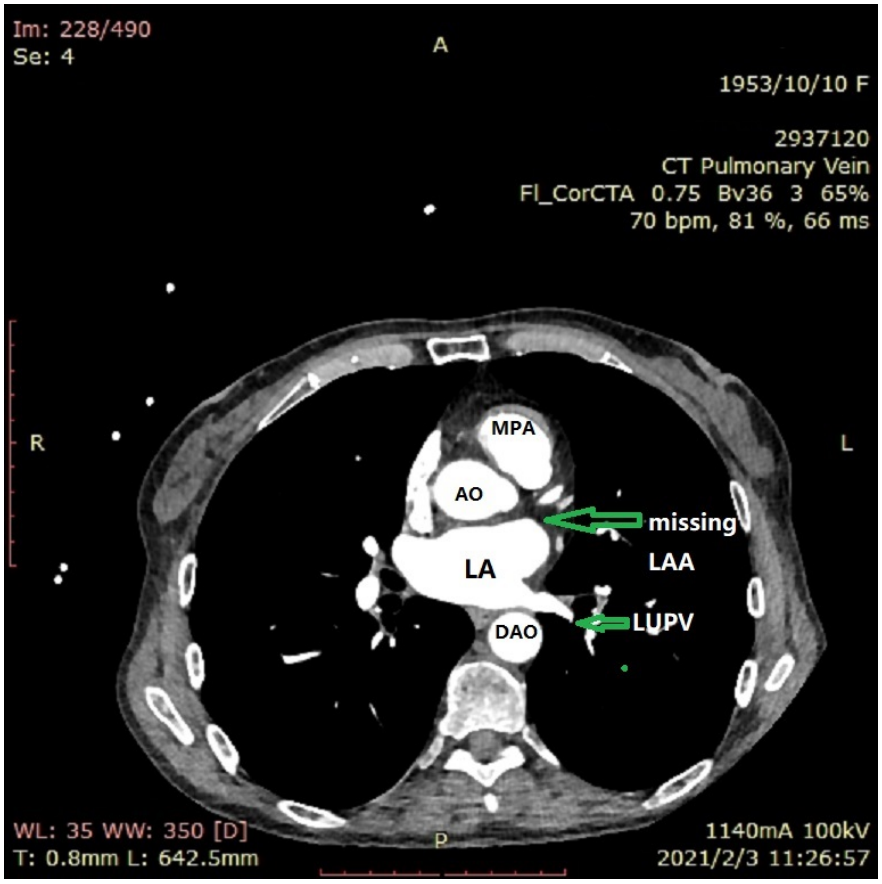
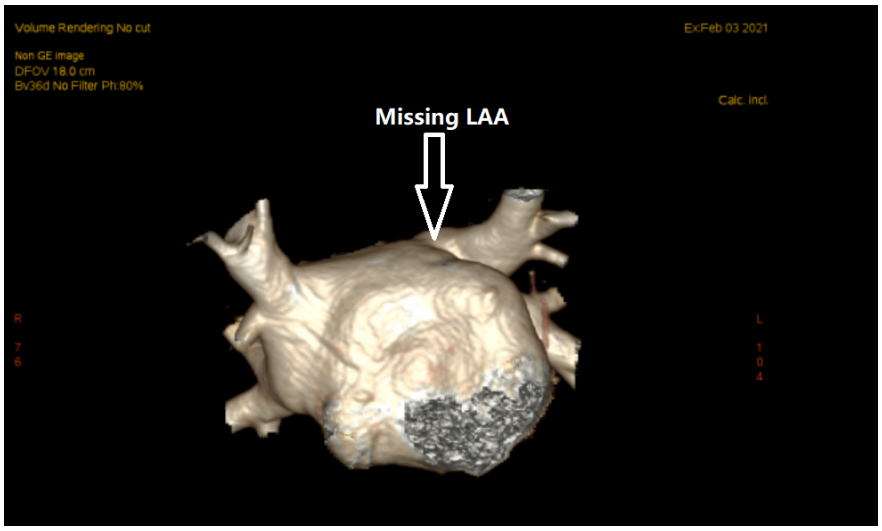


Figure 2: computed tomography of pulmonary veins showed no left atrial appendage





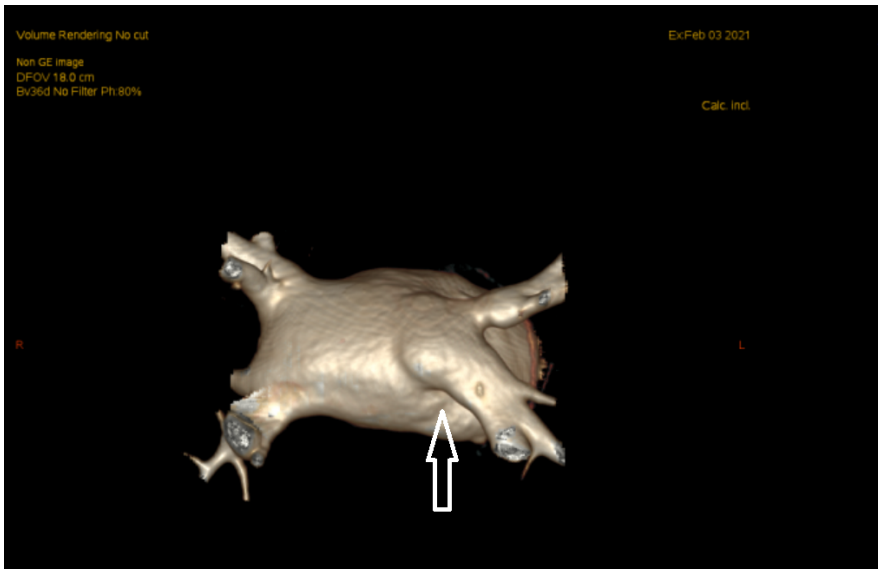


Figure 3: Plaque of carotid artery