Case Report

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2023.51.008145

Abnormal Voltage in Electro-Anatomical Voltage Mapping of Left Atrial Appendage Absence: Case Report and Review of the Literature

Pang Ling Pin[1], Deng Si Min[1], Wu Wen Bin[1], Zhong Tong Chun[1], Wen Wen[1], Wu Yuan Cong[1], Chen Cheng Di[1], Luo Peng[1], Liu Hao[2], Ba Ming Chuan[2], Guo Lin[2], Cai Fu Sheng[2], Li Min Fei[2], Xu Pei Yuan[2], Li Li Jun[2], Gao Zhen[2], Su Zi Ce[3], Huang Shi An[1]*, Zhang Hong Zhe[2]*

[1]*Department of Cardiology, Affiliated Hospital of Guangdong Medical University, Zhanjiang, Guangdong, China*

[2]*Department of Cardiology, the Seventh Affiliated Hospital of Southern Medical University, Foshan, China*

[3]*The Second Clinical Medical School, Southern Medical University, China*

***Corresponding author:** Huang Shi An, Department of Cardiology, The Affiliated Hospital of Guangdong Medical University, Zhanjiang, China

Zhang Hong Zhe, Department of Cardiology, The Seventh Affiliated Hospital, Southern Medical University, Desheng Road section 28, Foshan 528200, China

ARTICLE INFO

Received: July 06, 2023

Published: July 21, 2023

Citation: Pang Lingpin, Zhong Tongchun, Wen Wen, Wu Yuancong, Chen Chengdi, Luo Peng, Liu Hao, Ba Ming Chuan, Guo Lin, Cai Fu Sheng, Li Min Fei, Pei Yuan Xu, Li Li Jun, Gao Zhen, Su Zi Ce, Huang Shi An, Zhang Hong Zhe. Abnormal Voltage in Electro-Anatomical Voltage Mapping of Left Atrial Appendage Absence: Case Report and Review of the Literature. Biomed J Sci & Tech Res 51(4)-2023. BJSTR. MS.ID.008145.

ABSTRACT

The absence of left atrial appendage (LAA) is an extremely rare disease, which is usually diagnosed by chance in imaging examinations before atrial fibrillation surgery. Here, we report a rare case of left atrial appendage loss in a 67-year-old man who planned to undergo radiofrequency catheter ablation of atrial fibrillation. We observed LAA deficiency in transesophageal echocardiography (TEE) and further refined left atrial CTA imaging and cardiac magnetic resonance imaging (MRI) to confirm this diagnosis. Since there is no data on anticoagulation in patients with a missing left atrial appendage, we continue to use rivaroxaban anticoagulation per guidelines after successful radiofrequency catheter ablation. For the diagnosis of patients with a lack of left atrial appendage, it is necessary to use a variety of imaging methods to clarify the absence of a left atrial appendage. In this case report, we focused on the characteristics of electro-anatomical voltage mapping by the left atrial appendage absence, discussed the direct relationship between left atrial appendage absence and atrial fibrillation, and briefly reviewed the characteristics of 19 cases reported in the literature so far.

Keywords: Left Atrium; Absent Left Atrial Appendage; Atrial Fibrillation; Catheter Ablation

Abbreviations: CTA: Complete Left Atrial; MRI: Magnetic Resonance Imaging; LAA: Left Atrial Appendage, TEE: Transesophageal Echocardiography

Introduction

The left atrial appendage is the remnant appendage of the primitive left atrium during the embryonic period, located between the left upper pulmonary vein and the left ventricular free wall. It is the most common site of atrial thrombosis in the presence of atrial arrhythmias such as atrial fibrillation. Early intervention of atrial thrombosis (such as left atrial appendage occlusion and drug anticoagulation therapy) is the main means to prevent vascular embolism, but this requires more understanding and research on the structure and function of the left atrial appendage. The shape of the LAA is variable, but the absence of the LAA is very rare and is likely due to congenital anatomical variations. Here, we report a rare case of LAA absence in a 67-year-old patient and focus on the characteristics of LAA absence in electro-anatomical voltage mapping. In addition, we briefly reviewed the clinical characteristics of 19 cases with missing LAA.

Case Report

A 67-year-old male patient experienced dizziness, and palpitation, accompanied by nausea and vomiting 3 days before admission. He came to the emergency department of our hospital for treatment. ECG showed "atrial fibrillation with rapid ventricular rate, frequent multi-source ventricular premature beats, and paired phenomenon." He came to our department for further diagnosis and treatment. Previous history of hypertension, hyperlipidemia, impaired glucose tolerance, no history of cardiac-related surgery. The diagnosis of "1. Paroxysmal atrial fibrillation and 2. Frequent ventricular premature beats" was considered on admission. According to the patient's condition and willingness, the patient was selected to undergo radiofrequency catheter ablation under three-dimensional mapping [1-6]. After admission, NT-proBNP was 1048.000| pg/ml. Esophageal echocardiography was completed before surgery, and no thrombus or left atrial appendage was found (Figure 1). Although esophageal echo imaging was obtained at multiple acquisition angles, LAA imaging could not be acquired and the absence of LAA was suspected. To further confirm and clarify the diagnosis, the patient underwent complete left atrial CTA imaging (Figure 2) and cardiac magnetic resonance imaging (MRI) examination (Figure 3).

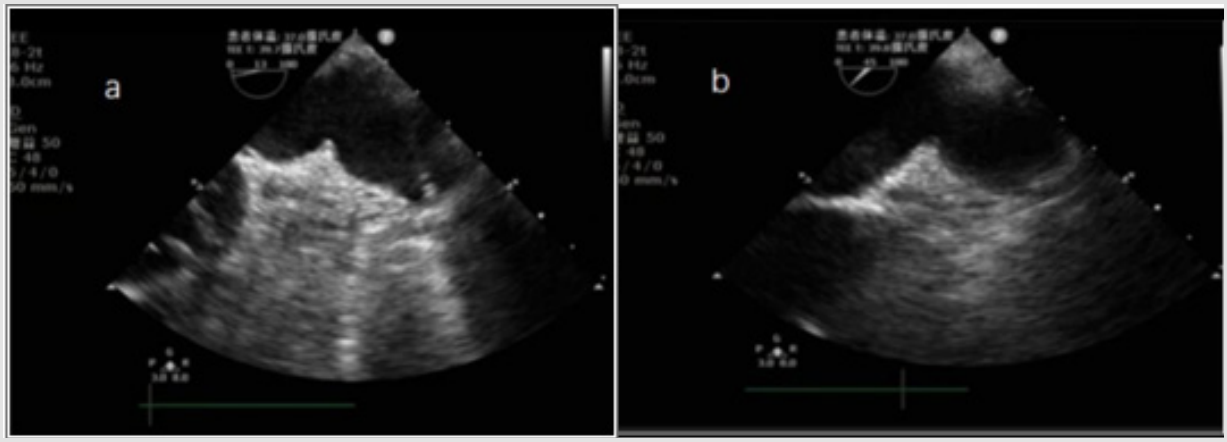

Figure 1: Transesophageal electrocardiographic imaging of the left atrium.

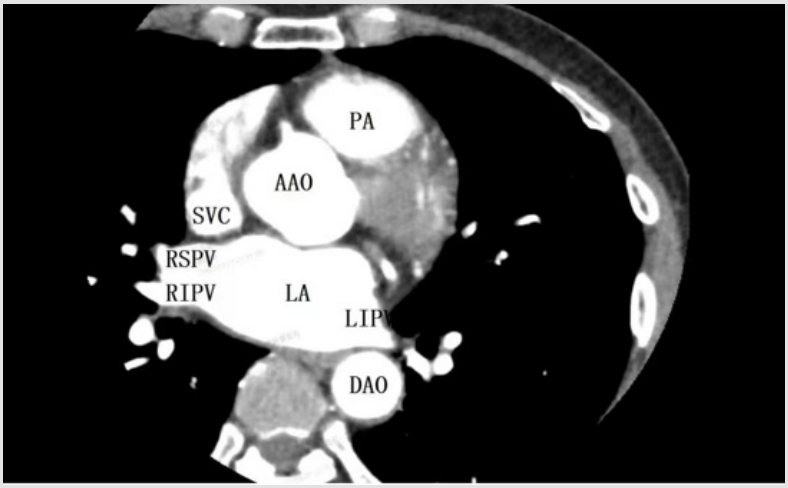

Figure 2: Complete left atrial CTA imaging.

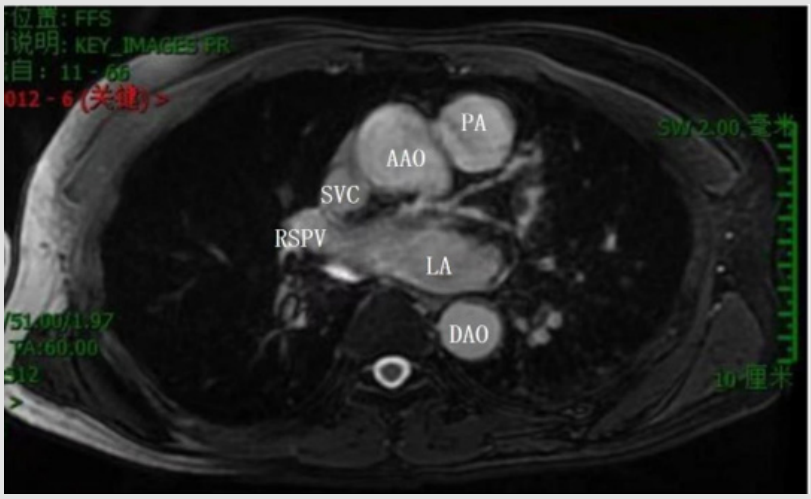

Figure 3: Cardiac magnetic resonance imaging (MRI) examination.

Both modalities confirmed the diagnosis of absent LAA and no intracardiac thrombus. On the day of surgery, we performed left atrial CTA/MRI and Carto-3 electroanatomical mapping system fusion, respectively, and reconstruction was performed (Figure 4). During the surgery, substrate mapping was performed with a pentaray catheter (Figure 5), and an abnormal voltage area higher than the adjacent atrial tissue was recorded at the original position of the left atrial appendage (Figure 6). Circumferential pulmonary vein electrical isolation was successfully achieved. During the surgery, potential disappeared and sinus rhythm was converted.No atrial arrhythmia was induced by repeated isoproterenol and high frequency (220ms and 200ms), and the procedure was successful. There are no data on anticoagulation therapy for atrial fibrillation with the absence of left atrial appendage, so rivaroxaban was given for anticoagulation therapy according to the CHA2DS2-Vasc score.

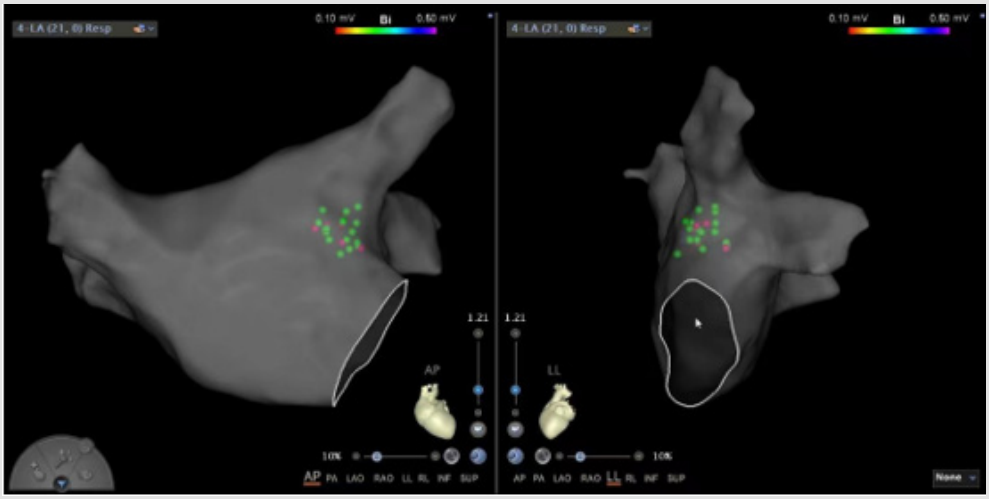

Figure 4: Left atrial CTA/MRI and Carto-3 electroanatomical mapping system fusion, respectively, and reconstruction was performed.

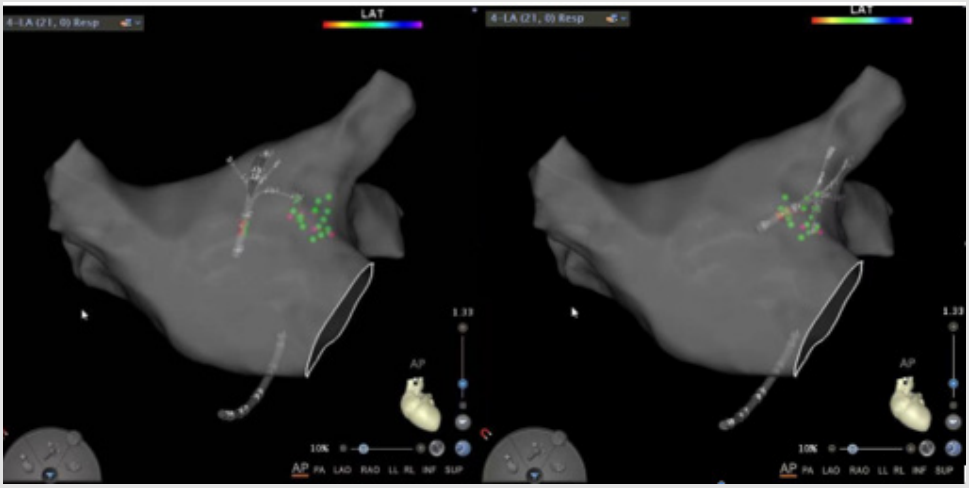

Figure 5: Substrate mapping was performed with a pentaray catheter.

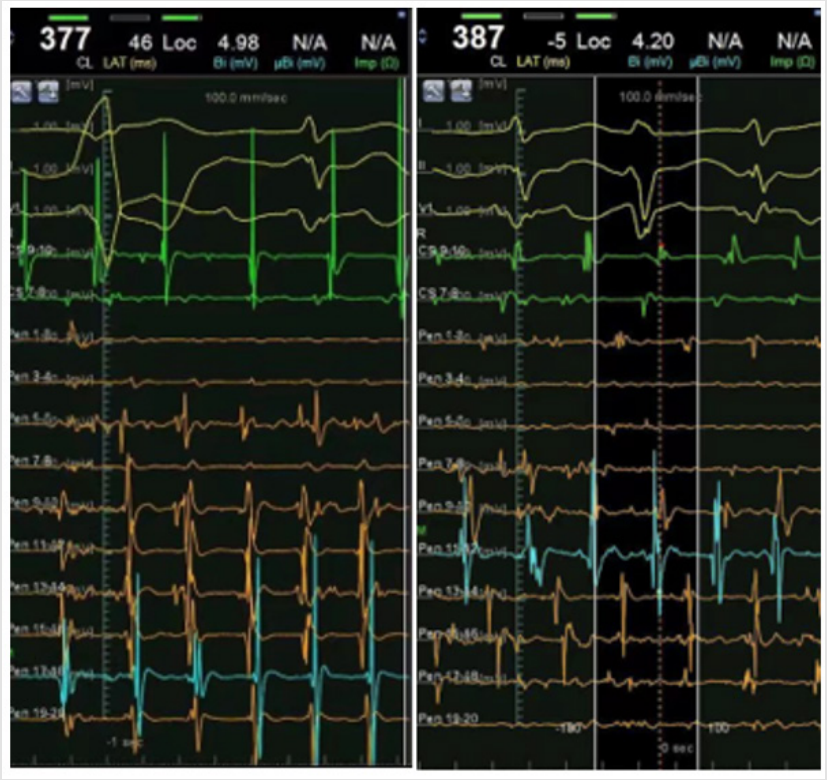

Figure 6: An abnormal voltage area higher than the adjacent atrial tissue was recorded at the original position of the left atrial appendage.

Discussion and a Literature Review

The left atrial appendage is usually located in front of the pericardial cavity, walking along the anterior wall of the left atrium, with the tip pointing in the direction between the pulmonary artery, right ventricular outflow tract, and left ventricular free wall. The edge is serrated, and there are abundant pectinate muscles and muscle trabeculae inside. The morphology of the LAA can be roughly divided into the following 4 categories: Windsock type, cauliflower type, chicken wing type, and cactus type. Absence of the LAA is rare and the exact prevalence and incidence of this condition are unknown. To our knowledge,

the first left atrial appendage deletion was reported by Collier et al since 2012,19 cases of which have been reported in the literature. Of the 20 reported cases, including ours, 8 were female (40%) and 11 were male (55%)(and gender was not mentioned in one report). In all patients, the abnormality was noted during preparation for AF ablation or during the assessment of the heart for thrombus. Of the 20 reported cases, including this one, the disease was identified in 14 patients (70%) during the pre-procedure assessment for AF ablation. Two patients (10%) were diagnosed in preparation for LAA occlusion. Two patients (10%) had a diagnosis made by esophageal echocardiography before cardioversion of AF. In 1 patient (5%) who had a stroke, the disease was identified on TEE at the time of embolus source assessment [7-13]. One patient (5%) was found on complete TEE before cardiac synchronization therapy. TEE has proven to be adequate in assessing LAA loss from the current case reports. However, the LAA may not be visible due to complete thrombus occlusion, rare anatomical features, previous relevant LAA procedures, and poor imaging quality.

In addition, multimodality imaging such as cardiac ct scans, cardiac magnetic resonance imaging, and atrial angiography is often required to confirm changes in the location and morphology of the left atrial appendage.

If the ostium is obstructed, the LAA may not be visible on left atrial angiography, but a combination of imaging modalities including TEE, cardiac CT, and LA angiography can provide an accurate diagnosis. Only sporadic cases of missing LAA have been reported in the literature, and identification of the disease usually requires advanced cardiac imaging of the patient, so it seems impossible to estimate the prevalence of the disease [14-20]. We have carried out the heart gene aspect research on this patient, obtains the related heart gene result, and detects RYR2; NM_001035.2:c.9797C>T(p.Thr3266Ile) mutation, there is no report on the pathogenicity of this mutation. According to the ACMG guidelines (Appendix), this variation was judged to be of undetermined significance. However, some studies have shown that this gene often leads to the occurrence of catecholamine-sensitive polymorphic ventricular tachycardia. A thrombus associated with atrial fibrillation is mainly formed in the left atrium. Currently, patients who are not suitable for anticoagulation are treated with left atrial appendage resection and left atrial appendage occlusion. Anticoagulation therapy in patients lacking LAA remains unclear. Although the risk of thrombosis is significantly reduced in patients with congenital absence of LAA, it does not disappear completely. Therefore, for patients without contraindications, anticoagulation should be initiated according to the guideline score to reduce the risk of thromboembolic events. However, for patients who cannot tolerate anticoagulation or are at significant risk of bleeding, clinicians should weigh the risk-benefit of anticoagulation without the LAA. The complex structure of the left atrial appendage, with several layers of muscle bundles arranged in different directions and extensive pectinate muscles, is easy to cause the formation of reentry, which leads to the electrical remodeling of the atrium and finally leads to the triggering of atrial fibrillation [21].

In addition, the histological origin of the LAA may be related to the cause of AF due to an ectopic pacemaker in the LAA. Douglas et al. found that the LAA-LAA junction is histologically similar to the CS, and hypothesized that the increased composition of the vessel wall during the entry of the PV and surrounding myocardium into the LA may reduce the entry of the sinus myocardium to the area surrounding the LAA, where slow conduction or conduction block may occur, perhaps as a prerequisite for inducing reentry. Our case report differs from other reports in that we used the carto system for substrate mapping with the pentaray catheter, which recorded the left atrial substrate and observed areas of abnormal voltage above the adjacent atrial tissue at the original location of the left atrial appendage. Electroanatomical voltage mapping has the potential to identify areas of atrial scar that may play a key role in AF substrates.

Although the etiology of atrial scarring has not been fully elucidated, previous reports have suggested that LA scarring is an independent predictor of long-term recurrence. To our knowledge, this is the first case report describing an atrial voltage-substrate map with abnormal voltages in a congenital absence of the LAA. Unlike Enomoto et al., who mapped the left atrial voltage map, they did not observe any abnormal potentials or low voltage areas in the LAA. Myocardium formation at the site of the missing LAA in this patient may be different from other cases, but these require further investigation of tissue formation in the associated atrium.

Conclusion

In the present report, we describe a 67-year-old man with a rare LAA diagnosed by multimodality Imaging, who described an atrial voltage substrate map with abnormal voltage in a congenital LAA absence and who also underwent gene sequencing. Through the review of the literature, we need to consider the congenital absence of the left atrial appendage when we encounter non-visualization of the left atrial appendage in clinical practice. Transesophageal three-dimensional echocardiography is usually sufficient for diagnosis, but cardiac CT and cardiac MRI are complementary imaging modalities. For patients with LAA loss and atrial fibrillation, anticoagulation should be initiated according to guideline scores to reduce the risk of thromboembolic events if there are no contraindications.

Acknowledgement

Construction of Zhanjiang Vascular remodeling and arrhythmia Medical Engineering Research Center.

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ISSN: 2574-1241

DOI: 10.26717/BJSTR.2023.51.008145

Zhang Hong Zhe and Huang Shi An. Biomed J Sci & Tech Res