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Congenital absence of left atrial appendage: Case report and review of the literature

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Abstract

Congenital absence of left atrial appendage (LAA) is a rare condition, mostly diagnosed incidentally during imaging examination intended for other purpose. We reported a case of congenital LAA absence diagnosed in a 55-years-old man by trans-esophageal echocardiography (TEE) and confirmed by computed tomography (CT). We also conducted a systematic review of the twenty-eight cases reported in the literature.

Keywords: Absent left atrial appendage; Left atrial appendage agenesis; Atrial fibrillation trans-esophageal echocardiography; Anticoagulant therapy.

Abbreviations: LAA: Left Atrial Appendage; LA: Left Atrium; TEE: Trans-Esophageal Echocardiography; CT: Computed Tomography; MESH: Medical Subject Heading; AF: Atrial Fibrillation; 3D: Three-Dimensional; ICD: Implantable Cardioverter Defibrillator.

Introduction

Left atrial appendage (LAA) is a finger-like muscular extension of the left atrium (LA). It acts as blood reservoir, modulates LA pressure and elicits adaptive responses to stress. Several LAA shapes and variants have been described. We report a rare case of congenital absence of LAA diagnosed by trans-esophageal echocardiography (TEE) and confirmed by computed tomography (CT) in a 55-year-old patient. Additionally, we describe the characteristics of this congenital anomaly by reviewing relevant literature.

Materials and Methods

A systematic literature searches of PubMed and Google Scholar databases for case reports and related articles of absence/agenesis of left atrial appendage (LAA) published until July 2023 was conducted using "left atrium", "appendage", "congenital", "atrial," and "absent" in addition to the medical subject heading

(MESH) terms “atrial appendage” as search terms. Twenty-three cases were identified among articles published. The characteristics of diagnostic modalities, demographic distribution and prognosis of this anomaly were studied.

Case Report

A 55-year-old male patient with history of hypertension, underwent a trans-esophageal echocardiogram for an undated episode of atrial fibrillation (AF). TEE showed normal LA volume without thrombi or echo contrast, but despite the three-dimensional acquisition, we were unable to identify LAA. Congenital absence of LAA was suspected and multi-detector cardiac CT confirmed the abnormality (Figure 1).

Results

In literature, 27 cases of congenital absence of LAA have been reported: the first case was described in 2012 by Collier et al. [1-24] among the 27 reported cases, 15 were men (53,6%) and 10 women (35,7%) (in 3 reports gender was not mentioned) with mean age of 63,3±16,7 years. Characteristics of the cases are reported in Table 1. 89% of the patients had a medical history of atrial fibrillation or atrial flutter.

In about 90% of cases, the abnormality was identified incidentally during imaging examinations performed before AF ablation, cardioversion procedures, LAA closure and implantable cardioverter defibrillator (ICD) implantation.

In 1 case the LAA absence was identified during a cardiac tomography performed for angor and dyspnea [19], in another case during TEE performed to evaluate intracardiac thrombi in a stroke patient [17] and in the last case during autopsy [11]. Only 5 patients had other congenital cardiac anomalies [4,11,18,21,22] but the embryological mechanism of this anomaly remains unknown. Only 3 patients had history of stroke and 2 compatibles with embolic process [7,17,24].

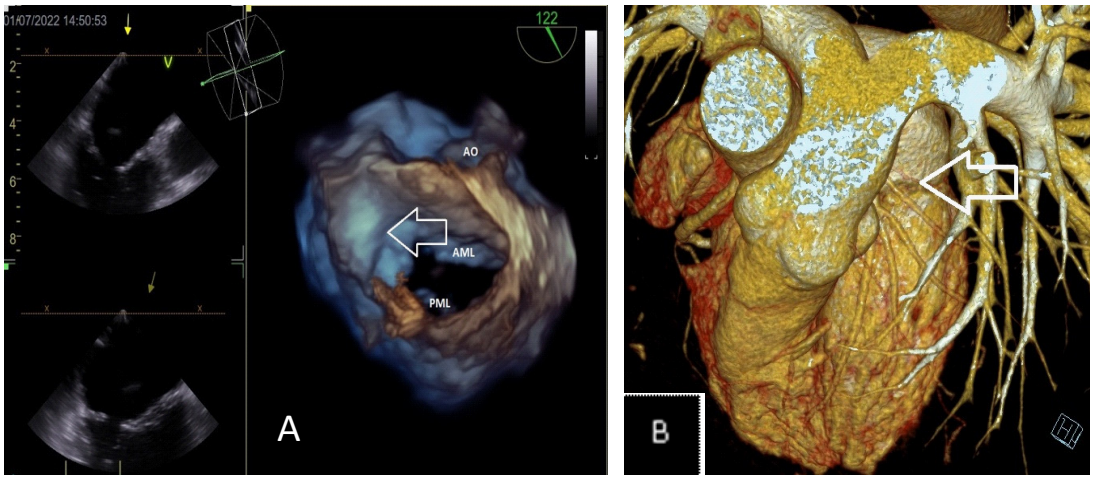

Figure 1: Congenital absence of LAA was suspected and multi-detector cardiac CT confirmed the abnormality.
A: 2D and 3D trans-esophageal echocardiogram: the arrow show absent of left atrial appendage.
B: Heart computer tomography. The arrow evidences no left atrial appendage.

Table 1: Characteristics of the cases are reported.

Article/Date	Gender (m/f)	Age (Years)	Past Medical History	Associated Congental Pathology	Diagnosed by	Diagnostic Study/ Studies Performed	Anticoagulant (NAO/TAO)
Collier [12] (2012)	F	73	AF	/	Incidental finding during preprocedural TEE for AF cardioversion	TEE	Unknown
Zhang [16] (2013)	M	60	HTN, DM, CAD, AF	/	Incidental finding during preprocedural TEE for AF ablation	TEE + 3D Reconstruction; Cardiac CT angiography; Left atrial angiography	Unknown
De ponti [4] (2014)	Unknown	52	Idiopathic paroxysmal AF	Persistent left SVC draining into the distal coronary sinus	Incidental finding during preprocedural TEE for AF ablation	TEE; Cardiac CT angiography	Unknown
Rosso [9] (2015)	M	62	Paroxysmal AF	/	Incidental finding during preprocedural TEE for AF ablation	TEE; Cardiac CT angiography; Left atrial angiography	Unknown
di Gioia [6] (2015)	M	78	HTN, HLP, AF, History of intracranial bleeding with warfarin	/	Incidental finding during pre-procedural evaluation of LAA closure	TEE; Cardiac CT angiography	No (Single antiplatelet therapy)
Ghori [8] (2015)	M	50	AF	/	Incidental finding during pre-procedural TEE for AF cardioversion	TEE; Cardiac CT angiography; Cardiac MRI	Yes
Saleh [15] (2015-1)	F	54	CAD-STEMI, CHF, AF	/	Incidental finding during preprocedural TEE for AF ablation	TEE; Cardiac CT angiography	Yes
Saleh [15] (2015-2)	M	79	HTN, AF, non-ischemic cardiomiopathy	/	Incidental finding during pre-procedural TEE for biventricular ICD insertion	TEE	Unknown
Katsumata [22] (2016)	F	76	HTN, paroxysmal AF	Persistent left SVC	Incidental finding during pre-procedural CT for AF ablation	TEE; Cardiac CT angiography; Left atrial angiography	Unknown
Song [24] (2016-1)	M	68	HTN, DM, CAD, AF	/	Incidental finding during pre-procedural CT for AF ablation	Cardiac CT angiography; Left atrial angiogram	Yes (Warfarin for 12 months after ablation)
Song [24] (2016-2)	F	58	HTN, lacunar stroke, AF	/	Incidental finding during pre-procedural CT for AF ablation	Cardiac CT angiography; Left atrial angiogram	Yes (Rivaroxaban)
Kureshi [21] (2017)	F	62	Obesity, HTN, AF	Single left sided and 2 right-sided pulmunary veins	Incidental finding during pre-procedural CT for AF ablation	Cardiac CT agiography; TEE; 3D CT reconstruction of the LA in electroanatomic mapping system	Unknown
Dar [10] (2017)	M	67	HTN, DM, AF	/	Incidental finding in pre-procedural CT for MAZE procedure for AF	Cardiac CT angiography	Unknown
Enomoto [5] (2018)	M	70	HTN, AF	/	Incidental finding during pre-procedural TEE/CT for AF ablation	TEE; Cardiac CT angiography; Left atrial angiography; 3D voltage mapping of LA	Yes but stopped anti- coagulation 6 months after AF ablation as there was no AF recurrence and absent LAA
Lee [17] (2018)	M	58	Type 2 DM	/	Work up for source of emboli	TTE; TEE; Cardiac CT angiography	Unknown

Nandar [23] (2018)	F	77	HTN, HLP, AF	/	Incidental finding during pre-procedural TEE/CT for AF ablation	TEE; Cardiac CT angiography	Yes (Warfarin)
Singam [1] (2018)	F	79	COPD, HTN, HLP, DM, CHF, persistent AF, intolerance for oral anticoagulation	/	Incidental finding during pre-procedural evaluation of LAA closure	TTE; TEE + 3D; Left atrial angiography; Cardiac CT angiogrqaphy	Yes
Pashun [19] (2019)	M	42	No past medical history, no AF	/	Incidental finding during CT angiography for angor and dyspnea	Cardiac CT angiography	Unknown
Pourafkari [3] (2020)	M	80	COPD, HTN, Atrial flutter	/	Incidental finding during pre-ablation evaluation	TEE; Cardiac CT angiography with 3D reconstruction	Yes (Rivaroxaban)
Vaideeswar [11] (2020)	M	0,25	History of respiratory distress	Ostium secundum atrial septal defect; Ventricular septal defect, Trasposition of the great vessels; Mitral atresia; Single ventricle of right ventricular morphology with a double outlet	Incidental finding during autopsy after death for adenoviral interstitial pneumonitis with superadded confluent bronchopneumonia	TEE; Complete autopsy	No
Mayire [7] (2021)	F	68	HTN, DM, History of ischemic stroke; paroxysmal AF (ablation refractoriness)	/	Incidental finding during pre-procedural evaluation of LAA closure	TEE + 3D Reconstruction; Cardiac CT angiography	Yes (Warfarin)
Li [2] (2022)	M	57	HTN, DM, paroxysmal AF (electric CV refractoriness)	/	Incidental finding during pre-ablation evaluation	TEE; Cardiac CT angiography	No
Meeks [20] (2022)	F	70	Paroxysmal AF	/	Incidental finding during pre-ablation evaluation	Cardiac CT agiography; TEE + contrast-enhanced imaging acquisition (i.v. Perflutren Lipid Microspheres - Definity)	Yes
Vartak [14] (2023)	F	92	Atrial flutter, asthma, other comorbidities unknown	/	Incidental finding during pre-procedural TEE for atrial flutter cardioversion	TEE; Cardiac CT angiography	Unknown
Arguelles [18] (2023 -1)	Unknown	57	Paroxysmal AF, apical hypertrophic cardiomyopathy; ICD implantation after VT	Anomalous right coronary artery originating from the left coronary cusp through a slit-like ostium with an interarterial course	Incidental finding during preprocedural TEE for AF cardioversion	TEE + 3D Reconstruction; Cardiac CT angiography	Yes (Rivaroxaban)
Arguelles [18] (2023 -2)	Unknown	71	Paroxysmal AF, HLP, severe meta-rheumatic mitral regurgitation awaiting for surgical MVR	/	Incidental finding during pre-procedural evaluation of LAA closure	TEE; surgical visualization	Unknown
Saghir [13] (2023)	M	58	Paroxysmal AF, CAD, DM, CHF, anticoagulation intollerance	/	Incidental finding during pre-procedural evaluation of LAA closure	TEE; Cardiac CT angiography	Unknown
Current (2022)	M	55	HTN, AF	/	Incidental finding during pre-procedural TEE for AF cardioversion	TEE + 3D Reconstruction; Cardiac CT angiography	Yes

(AF: Atrial Fibrillation; CAD: Coronary Artery Disease; CHF: Congestive Heart Failure; COPD: Chronic Obstructive Pulmonary Disease; CT: Computed Tomography; CV: Cardioversion; DM: Diabetes Mellitus; F: Female; HLP: Hyperlipidemia; HTN: Hypertension; ICD: Implantable Cardioverter-Defibrillator; LA: Left Atrium; LAA: Left Atrial Appendage; M: Male; MPR: Multi-Planar Reconstruction; MRI: Magnetic Resonance Imaging; MVR: Mitral Valve Replacement/Repair; STEMI: ST Elevation Miocardial Infarction; SVC: Superior Vena Cava; TEE: Transesophageal Echocardiography; TTE: Transtoracic Echocardiography).

Discussion

Left atrial appendage (LAA) is a finger-like muscular extension of the left atrium (LA) located close to the left ventricular free wall [25] and close to the left circumflex artery [26]. Several shapes and sizes of the LAA were described [27] and multiple imaging modalities can be used to define the its anatomy or to detect any in situ thrombosis, but the most widely used technique is TEE, especially with 3D reconstruction system [21]. As we showed, LAA agenesis is a rare condition but its real prevalence and incidence remains unknown because it is always an occasional finding. It seems more frequent in male patients and it can be associated with congenital heart defects that we should always look for. The role of this abnormality in thrombotic risk assessment has not been studied, but it is known that in patients with non-valvular AF, about 10% of intracardiac thrombi form at sites other than the LAA [28], so in these patients, the thrombotic risk remains no negligible. The congenital absence of LAA could be also considered similar to LAA percutaneous occlusion or surgical exclusion which are less effective than anticoagulants (IIb class of recommendation in ESC guideline Atrial Fibrillation 2020) [29]. Therefore, in our opinion, anticoagulant therapy should be considered, based on the traditional risk factor approach, using the clinical risk score for stroke CHA2DS2-VASc.

Conclusion

Congenital absence of LAA is an extremely rare condition, and despite reported cases are increasing, its embryological origin and its pathophysiological role in thromboembolic risk, is still not well defined.

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