CASE REPORT

Congenital absence of left atrial appendage: A case report and literature review

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Congenital absence of left atrial appendage (LAA) is an extremely rare entity. A 50-year-old man with no past cardiac history was admitted with symptomatic atrial fibrillation (AF). Before subjecting him to direct current (DC) cardioversion, the patient underwent a transesophageal echocardiography (TEE) examination, but neither LAA nor a cardiac thrombus could be detected. Absence of both was confirmed by cardiac computer tomography (CT) and a cardiac magnetic resonance imaging (MRI) scan. The patient reverted to sinus rhythm with an uneventful synchronized biphasic DC shock of 100 joules.

Keywords: Left atrium, Left atrial appendage, Atrial fibrillation

Introduction

Congenital absence of left atrial appendage (LAA) is an extremely rare cardiac anomaly. To date, only few cases have been reported [1,2]. We report this case as one of the rare findings on cardiac imaging studies as it might be of interest for physicians and echocardiographers.

Case summary

A 50-year-old man, with no prior cardiac history, presented with his first ever symptomatic episode of atrial fibrillation (AF). As part of the pre-procedure evaluation for an elective direct current (DC) cardioversion, the patient underwent transesophageal echocardiography (TEE) to exclude LAA thrombus. Despite acquiring good quality standard and modified views, the operators were unable to discern LAA or any intracardiac thrombus. (Fig. 1A–D). However, the patient was not cleared for DC cardioversion, as complete occlusion of LAA with iso-echogenic thrombus remained a possible reason for its invisibility. Congenitally absent or rudimentary LAA was another, albeit rare, likelihood. For further delineation, and taking into account the devastating effects on a patient's quality of life should an LAA thrombus be missed, the patient had contrast-enhanced multidetector cardiac computed

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tomography (CT) and cardiac magnetic resonance imaging (MRI). Both of these modalities confirmed a diagnosis of congenitally non-existing LAA as well as no intra-cardiac thrombus (Figs. 2 and 3). The patient was subjected to a successful DC cardioversion under cover of a recommended anticoagulation regime.

Abbreviations

TEE	transesophageal echocardiography
LAA	left atrial appendage
AF	atrial fibrillation
MRI	magnetic resonance imaging
DC	direct current

Discussion

We report this case to highlight a rare congenital cardiac anomaly of unknown physiological consequence. LAA is a small, muscular extension abutting from the upper part of the left atrium. Typically developing in the third week of embryonic life [3], in adults it is described as a narrow, tubular, single or multi-lobed structure functioning as a decompression chamber during left ventricular systole and other periods of high left atrial pressure [3].

Considered a benign structure otherwise, LAA has high tendency to hatch a thrombus in patients with atrial fibrillation and with low flow states [3]. Such a clot may embolize peripherally, resulting in ischemic insult to the brain, kidneys and other organs supplied by the systemic circulation [4]. It is therefore imperative to ensure the absence of any LAA thrombus before a patient with AF can be cleared for DC cardioversion.

Advanced sonographic techniques such as biplane and multiplane TEE have enabled physicians to visualize LAA in most cases. A non-visualized LAA should, however, prompt the physician to consider possibilities such as indistinguishable complete thrombotic occlusion of the appendage, its prior percutaneous or surgical ligation, poor echocardiographic windows or a congenitally absent LAA, as in this index case. For further differentiation, highly sophisticated imaging modalities like multidetector CT and cardiac MRI would be of immense help, as demonstrated in our patient's case.

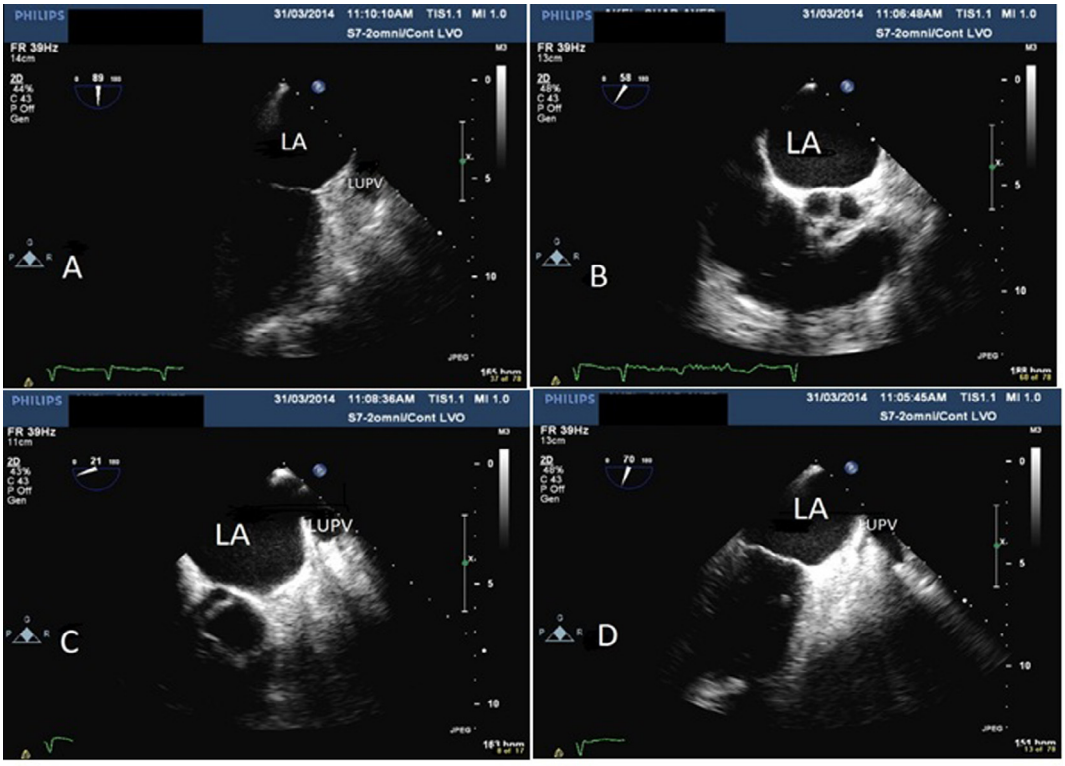

Figure 1. Transesophageal electrocardiographic imaging of the left atrium. Multiple views were obtained (A–D) at midesophageal level, but it was not possible to visualize the left atrial appendage. LA indicates left atrium. LUPV indicates Left upper pulmonary vein.

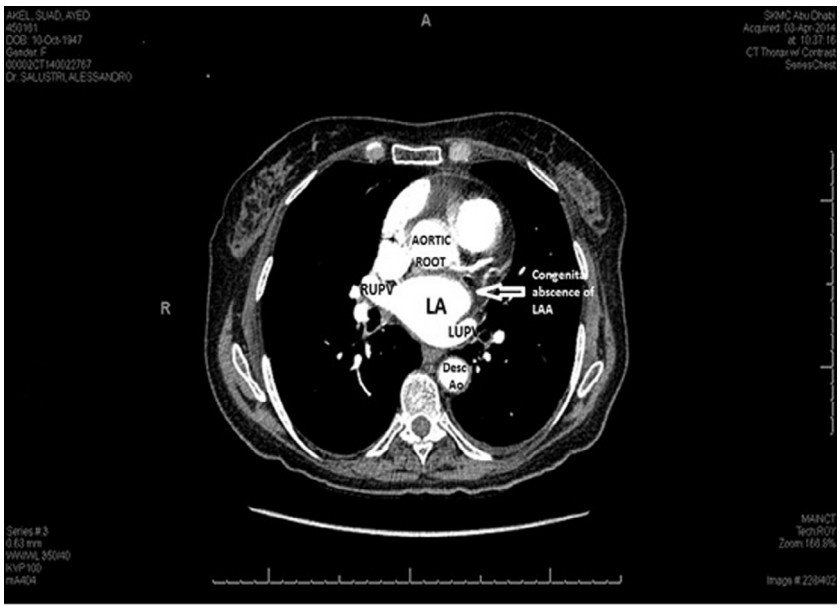

Figure 2. Multidetector computed tomography axial image of the left atrium depicting absence of LAA at its normal position (left arrow). LAA indicates left atrial appendage; RUPV, right upper pulmonary vein; LLPV, left lower pulmonary vein; LV, left ventricle; GCV, great cardiac vein; AscAo, ascending aorta; DescAo, descending aorta; PA, pulmonary artery.

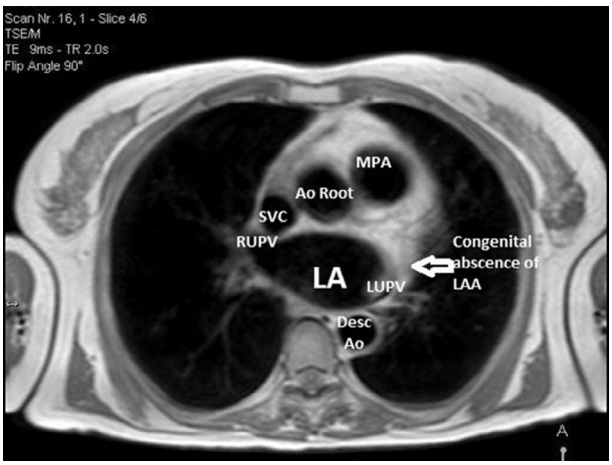

Figure 3. Axial Black blood cardiac Magnetic Resonance Image at the level of LA. Left arrow indicating absent LAA at its normal location. LA indicates left atrium, LAA, left atrial appendage; RUPV, right upper pulmonary vein; LLPV, left lower pulmonary vein; LV, left ventricle; GCV, great cardiac vein; AscAo, ascending aorta; DescAo, descending aorta; PA, pulmonary artery.

Conclusion

Congenital absence of LAA is an extremely rare cardiac anomaly and only a few cases have been reported so far. The authors believe that visualizing LAA by TEE may at times be challenging. Risk-free cardiac CT and MRI may confirm diagnosis and enable competent patient management.

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