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CASE REPORT

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Fenestrated membrane of the left atrial appendage orifice; an incidental finding of a rare congenital anomaly

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Abstract
Membrane of the left atrial appendage (LAA) is a rare congenital anomaly, which is reported in few cases, to date. Here, we reported an incidental finding of LAA obstructive membrane in patients with previous history of cerebrovascular event and newly diagnosed rheumatismal mitral stenosis (MS) in echocardiography.

Keywords Left atrial appendage, Fenestrated membrane, Rheumatismal mitral stenosis

Introduction

The left atrial appendage (LAA) is a little pouch located in the left atrium with a compound morphology leading to tendency to blood stasis, as it is the most prevalent locus of clot formation in patients with atrial flutter or atrial fibrillation [1]. Its orifice is in the anterolateral wall between the left upper pulmonary vein and the mitral annulus, just above the left circumflex artery. Close anatomic relationship of LAA to the free wall of the left ventricle distinguishes its anatomic characteristics from the left atrium [2]. The complex embryologic development causes a wide range of morphological variants and sometimes abnormalities might involve the appendage orifice. One of these abnormalities including membranes, which could prevent the pouch emptying, and predisposing the LAA to clot formation [3]. There are scarce reports of LAA membrane and its clinical importance is not understood well. Here, we reported the first case of concurrent rheumatismal mitral stenosis and LAA membrane.

Case presentation

A 64-year-old man presented with progressive cognitive symptoms together with ataxia and gate disturbance over the perior two months. He had a history of seizure from 30 years ago and right hemiplegia for one year. On admission, he had new left hemiplegia, dysarthria, urine incontinency and encopresis. He had exertional dyspnea during the last year without any chest pain. His vital sign was normal (Blood pressure: 110/70 mmHg, HR: 78 beats/min, T: 37.1 °C, SPO2: 96%, RR: 18/min). Patient has no positive familial history of similar conditions, cardio-embolic or cerebrovascular event and dysrhythmias, according to available data by taking history. In socio-medical history, he was opium addict from 10 years ago and consumed aspirin and sodium valproate. Brain magnetic resonance imaging (MRI) was indicative of diffused focal periventricular lesions and restriction in diffusion-weighted imaging (DWI), especially behind the corpus callosum. Moreover, periventricular and occipital enhancements were detected. Cerebrospinal fluid analysis had normal findings. Polymerase chain reaction

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tests (PCR) for tuberculosis, Epstein-Barr virus (EBV), human herpes virus (HSV), cytomegalovirus (CMV) and sarcoidosis had negative results. After excluding infectious and hemato-oncologic disorders, he underwent complementary assessments. CNS lymphoma was first suspected.

During patient re-evaluation and in more detailed physical examination, he had III/VI diastolic rumble in apex on heart auscultation. There was no edema in the lower limbs and pulse palpation was normal. Electrocardiogram had LA abnormality changes.

For determination of structural heart disease, he underwent transthoracic echocardiography (TTE) (Philips Healthcare, Andover, MA, The USA), and revealed normal biventricular size and function and rheumatic involvement of mitral valve leading to mitral stenosis. Because of very poor echo window, trans-esophageal echocardiography (TEE) was scheduled to precisely evaluate the severity of mitral stenosis and probable concurrent structural abnormalities. TEE showed significant left atrium (LA) enlargement, and severe LA smoke (sludge was noted in the left atrial appendage (LAA) associated with fresh clot formation at the bottom of LAA) (Fig. 1). A linear echogenic and semi-mobile structure was noted extending across the orifice of the LAA, with to-and-fro turbulent flow and relatively high-velocity jet (systolic velocity: 1.4 m/s; diastolic velocity: 1.2 m/s at the level of LAA orifice), suggestive of a fenestrated congenital obstructive membrane of the LAA (Fig. 2). Rheumatismal mitral valve (MV), up to moderate mitral regurgitation (MR), very severe mitral stenosis (MS) (MVA by 3D TEE: 0.65 cm2) and moderate aortic regurgitation were also reported. He was candidate for mitral valve (MVR) and aortic valve replacement (AVR) with LAA closure.

Discussion

The clinical importance of LAA membranes is not yet well known. It is very rare and less than 20 cases worldwide have been reported. To the best of our knowledge, this is the first case of LAA membrane concurrent with rheumatismal MS. Here, we reported an old man with previous CVA (cerebrovascular accident) and seizure and cardiac murmur who had rheumatismal MS in echocardiography together with incidental obstructive fenestrated LAA membrane.

The LAA membranes could be obstructive leading to functional stenosis when they are located at the ostium of the LAA, or non-obstructive if located in the body of the LAA. In our case, it was obstructive type. In two previous cases [4–6], obstructive membranes at the orifice of the LAA, leading to functional stenosis, were described and in three reports, non-obstructive membranes located in the body of the LAA were explained [6]. In another report, in a 26-year old patient, a partially obstructive membrane at the orifice of the LAA, causing functional stenosis was presented with no thrombosis [2]. Furthermore, Cresti et al. reported six patients with LAA membranes in a retrospective population-based study and announced that only one of them was obstructive associated with severely hypoplastic LAA. One patient had

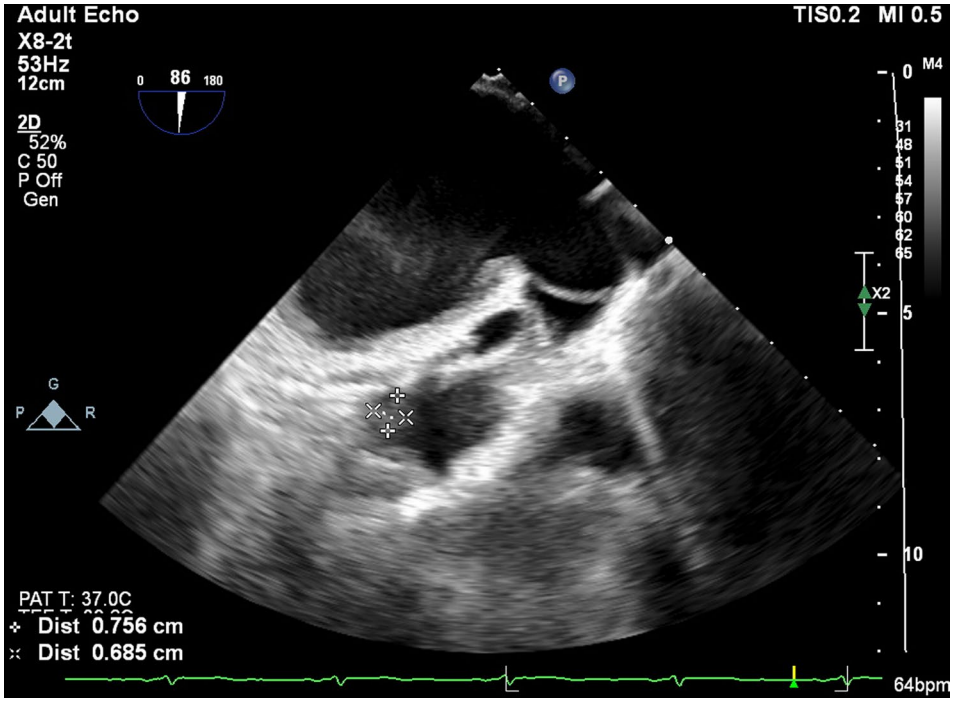

Fig. 1 TEE image of LAA; thrombosis is visualized at the bottom of LAA. TEE = trans-esophageal echocardiography, LAA = left atrial appendage

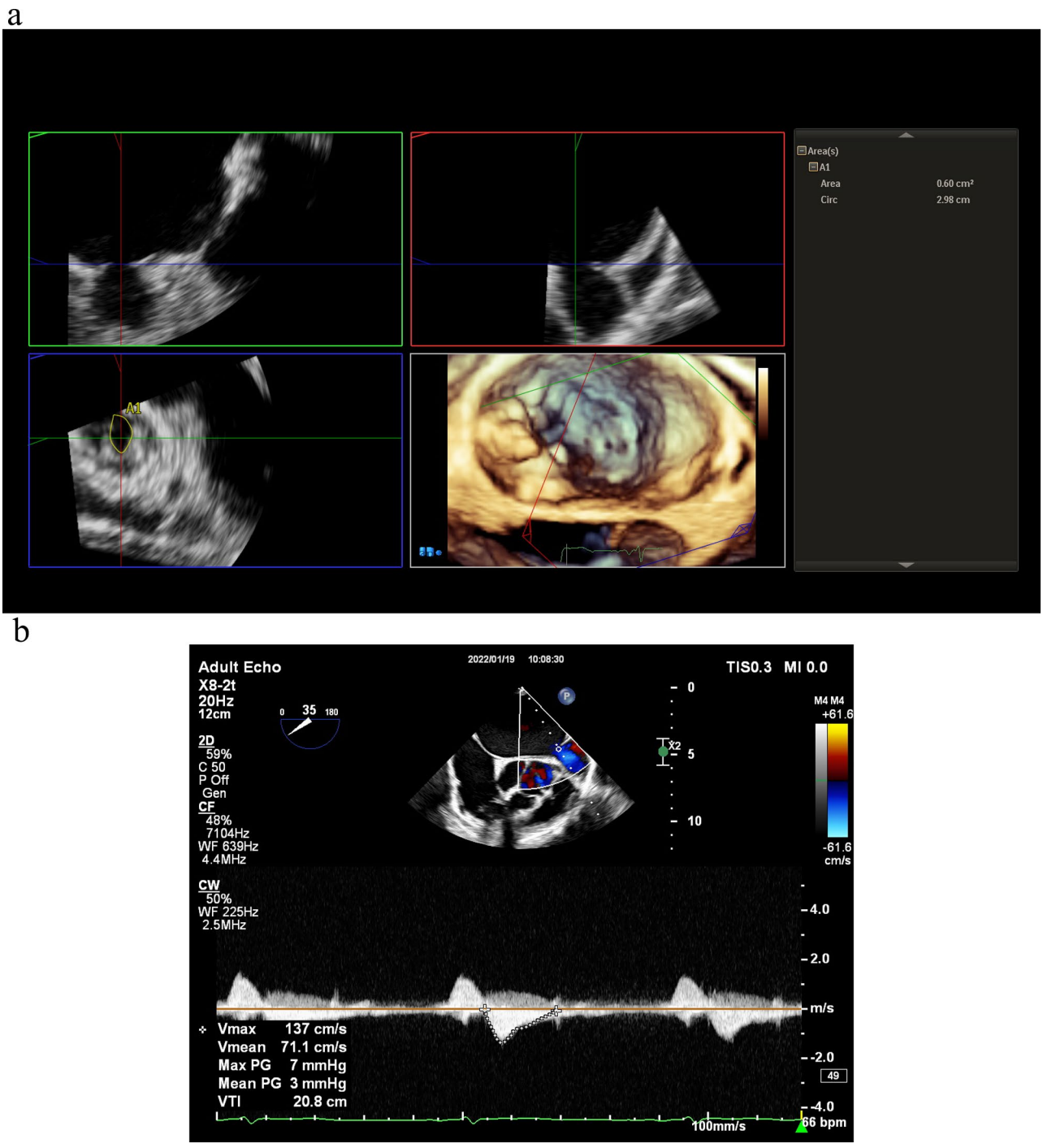

Fig. 2 TEE images; **a**) simultaneous multiplane imaging of LAA shows a linear echogenic structure extending across the orifice of the LAA; **b**) high-velocity jet with systolic velocity of 1.4 m/s and diastolic velocity of 1.2 m/s at the level of LAA orifice in CW Doppler study. TEE = trans-esophageal echocardiography, LAA = left atrial appendage

partially obstructive membrane with thrombosis formation [3]. Moreover, Correlae et al. reported one non-obstructive LAA membrane with no thrombosis [7]. On the other hand, Marinescu et al. described a partially obstructive fenestrated congenital membrane of the LAA without any thrombosis [8]. As it can be seen, there is no pattern in the obstructive or non-obstructive nature of the membranes.

In our patient, AF was not evident in ECG at the time of admission or during TEE. However, we could not exclude paroxysmal AF before this admission. In previous reports, AF or atrial flutter was an important finding. In

previous studies, most patients had atrial dysrhythmias [4–6]. In a recent study, mentioned above, it was shown that all six patients had AF rhythm (permanent, persistent or paroxysmal type) [3]. Despite a negative history of AF in our study, in case of incidental LAA membrane, atrial dysrhythmia should take in to account. Nonetheless, our patient had a history of repeated strokes which could be due to a transient arrhythmia. Finally, our patient underwent AVR, MVR and LAA closure. In another report, one patient had been presented with a transient ischemic attack [4].

Besides, there are some cases of failed surgical ligation of the LAA that were discovered to have a functionally stenosis of the LAA ostium due to incomplete ligation found during work up of atrial arrhythmia after surgery [9]. The physiology of an LAA membrane is similar to failed surgical LAA ligation or appendage trans-catheter closure device leaks, which may result in a functional stenosis of the LAA orifice. Consequently, the blood stasis may promote clot formation, but in these situation, clots could be less embolized due to the narrow residual orifice of LAA [6]. In our study, we found that thrombosis was made in LAA. In only one previous case, the thrombosis was reported as well [3]. However, due to small number of studies, cardio-embolic events could not be attributed to LAA membrane purely with high confidence.

Conclusion

This was the first case report without any documented AF rhythm and concurrent rheumatismal MS and obstructive LAA membrane. We explained a rare congenital anomaly, incidentally found in TEE which could not be easily assessed by other imaging modality like TTE. This condition might play a role in promoting atrial arrhythmia, affecting cardio-embolic risk or cerebrovascular events and complicating some catheter-based interventions.

Abbreviations

LAA	Left atrial appendage
MS	Mitral stenosis
MRI	Magnetic resonance imaging
PCR	Polymerase chain reaction
EBV	Epstein-Barr virus
HSV	Human herpes virus
CMV	Cytomegalovirus
TTE	Transthoracic echocardiography
TEE	Trans-esophageal echocardiography
LA	Left atrium
MV	Mitral valve
MR	Mitral regurgitation
MVR	Mitral valve replacement
AVR	Aortic valve replacement
CVA	Cerebrovascular accident

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Author contributions

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Not applicable.

Competing interests

The authors declare no competing interests.

Consent statement

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Consent to publish

Patient had consent to publish his data, anonymously.

Clinical trial number

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