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CASE REPORT
Cardiovascular imaging

Giant aneurysm of the left atrial appendage: a case report

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Background Left atrial appendage aneurysm (LAAA) is a rare condition mostly due to congenital malformations or secondary causes (i.e. mitral regurgitation).

Case summary We present a case of a 47-year-old male with a history of atrial fibrillation treated with propafenone presented to our emergency department for palpitation and epigastric pain. The electrocardiogram showed atrial fibrillation at high ventricular rate and a new-onset left bundle branch block. Urgent coronary angiogram excluded coronary artery disease. Echocardiography and cardiac magnetic resonance revealed a giant LAAA. The electrocardiogram alterations were deemed secondary to aberrancy and treatment with class IC antiarrhythmic. The patient was discussed in the heart team, and considering his will to avoid surgery, he was managed conservatively with closed follow-up, anticoagulant and antiarrhythmic therapy, and internal loop recorder. At 1-year follow-up, he showed asymptomatic and without arrhythmias.

Discussion Few cases are described in the literature; therefore, there is uncertainty in treatment and prognosis. Diagnosis is achieved with multimodality imaging. Treatment can be surgical with aneurysmectomy or conservative with regular follow-up by imaging examinations and pharmacological therapy aimed to prevent complications such as thrombosis and arrhythmias. Since high-quality scientific data are lacking, shared decision-making is essential for the management of patients affected by LAAA. In our clinical case, our patient's will to not undergo surgery was considered, and therefore, a conservative management with strict follow-up and medications was chosen.

Keywords Left atrial appendage aneurysm • Atrial fibrillation • Echocardiography • Cardiovascular imaging • Cardiac magnetic resonance • Case report

ESC curriculum 2.2 Echocardiography • 2.3 Cardiac magnetic resonance

Learning points

- Left atrial appendage aneurysm (LAAA) is a condition that can be highly symptomatic and can have serious complications.
- The main complications are intracardiac thrombi, arrhythmias, or compression of extracardiac or cardiac structures. If epicardial coronary arteries are compressed by the aneurysm, the patient can experience anginal symptoms and even myocardial infarction.
- The diagnosis of LAAA must comprehend transoesophageal echocardiography and other imaging modalities like cardiac computed tomography and cardiac magnetic resonance imaging to evaluate potential complications.
- Managing these patients can be challenging. Choosing between medical therapy in preventing complications or a surgical approach with aneurysmectomy is the main challenge; we therefore recommend a tailored approach based on every patient's needs and characteristics. We adopted a conservative approach after a collaborative evaluation of our heart team also considering our patient's will.

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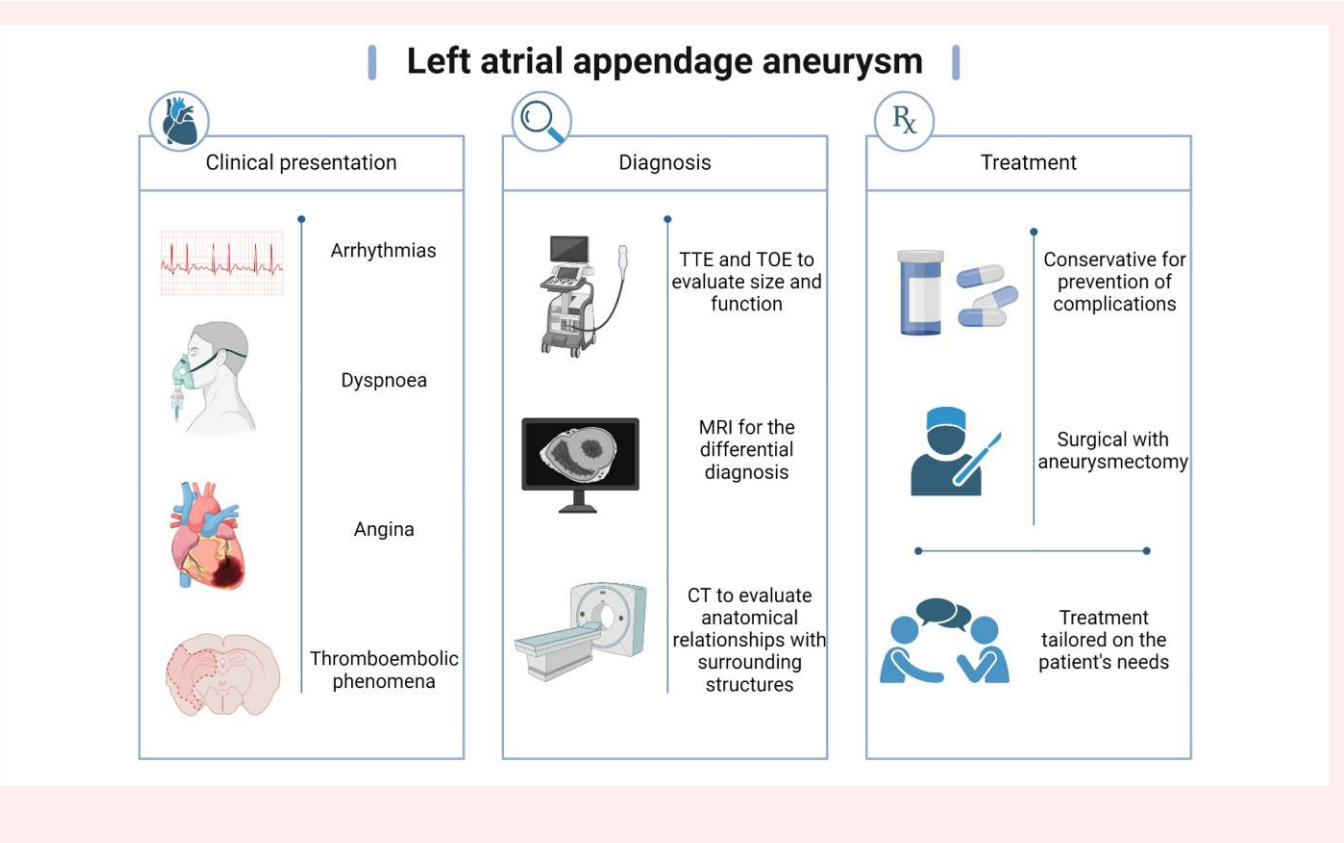
Introduction

Left atrial appendage aneurysm (LAAA) is a rare condition, usually acquired or congenital.[1,2] Its clinical presentation can be either asymptomatic or characterized by many signs and symptoms, including major cardiac or cerebrovascular events. The clinical presentation can vary from palpitations and concomitant dyspnoea or chest pain to supraventricular arrhythmias such as atrial fibrillation or flutter. Thrombotic formations and embolic phenomena may be the first clinical manifestation.[3]

To this date, <100 cases have been described, including paediatric ones. There is no standard of care for patients affected by this condition; therefore, patient-based management is usually required.

Summary figure



In this figure, we summarize the key steps to treat these patients. Left atrial appendage aneurysm is a cardiac malformation that can have a heterogeneous presentation with different signs and symptoms. Diagnosis is achieved through multimodality imaging: echocardiography (both transthoracic and transoesophageal) to assess the size and function of the cardiac chambers and possible complications such as intra-atrial thrombi, cardiac magnetic resonance imaging is useful in the differential diagnosis with other extracardiac masses of similar appearance on echocardiography, and cardiac computed tomography is useful in assessing anatomic relationships with surrounding structures. Treatment can be conservative with medical therapy to prevent complications or surgical with aneurysmectomy. There is insufficient evidence on the outcomes of different treatments, so individualized treatment is recommended according to each patient's needs.

Case presentation

A 47-year-old man was admitted to the emergency department for palpitations and epigastric pain. He had a history of paroxysmal atrial fibrillation, had previously undergone two pharmacological cardioversions, and had a history of mild arterial hypertension for which he was not taking any medication. At physical examination, he showed no signs congestive heart failure and irregular tachycardia of heart sounds without murmurs.

His electrocardiogram (ECG) on admission (*Figure 1A*) showed high-rate atrial fibrillation (150 b.p.m.) and new left bundle branch block. His echocardiogram showed left ventricular dysfunction with ejection fraction of 35% (see Supplementary material online, *Video S1*). An urgent coronary angiography was performed and found no significant coronary obstructions. Laboratory exams [high-sensitivity troponin I and N-terminal pro-brain natriuretic peptide (NT-proBNP)] were in the normal range.

At this point, the patient underwent a pharmacological cardioversion with amiodarone (5 mg/kg in 1 h and 50 mg/h for 24 h) and sinus rhythm was restored. The ECG acquired in sinus rhythm (*Figure 1B*) did not show the left bundle branch block, which was therefore attributed to a conduction aberrancy concomitant to the use of class IC antiarrhythmic drug propafenone, which the patient was taking at home.

A second transthoracic echocardiogram was performed once sinus rhythm had been restored, and it showed improvement of left ventricular systolic function (ejection fraction 57%). Furthermore, it showed a hypokinetic mid-apical segment of the anterolateral wall, corresponding to the site of a giant left atrial appendage aneurysm (*Figure 2*). To define better the LAAA, a transoesophageal echocardiogram was performed (*Figure 2*). It confirmed the aneurysmatic dilatation of the left appendage which appeared to have an oval-shaped neck (22 × 17 mm). No thrombotic

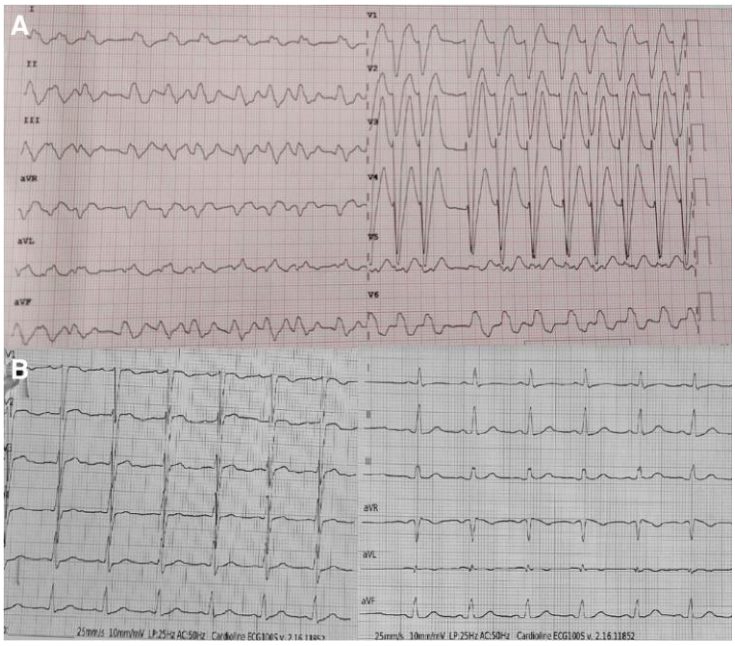


Figure 1 Electrocardiogram. (*A*) Atrial fibrillation with wide QRS during antiarrhythmic therapy with class IB drug. (*B*) Sinus rhythm and normal atrioventricular conduction after pharmacological cardioversion.

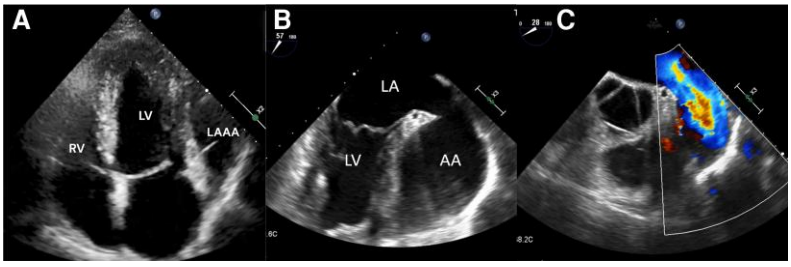


Figure 2 Echocardiography. (*A*) Transthoracic apical four-chamber view with the left atrial appendage aneurysm (LAAA) compressing the left ventricle lateral wall. (*B*) Mid-oesophageal two-chamber view showing the LAAA in its entire dimensions and showing no thrombi inside. (*C*) Mid-oesophageal short-axis colour Doppler view shows high velocities inside the auricle. LA, left atrium; AAA, atrial appendage aneurysm; LV, left ventricle.

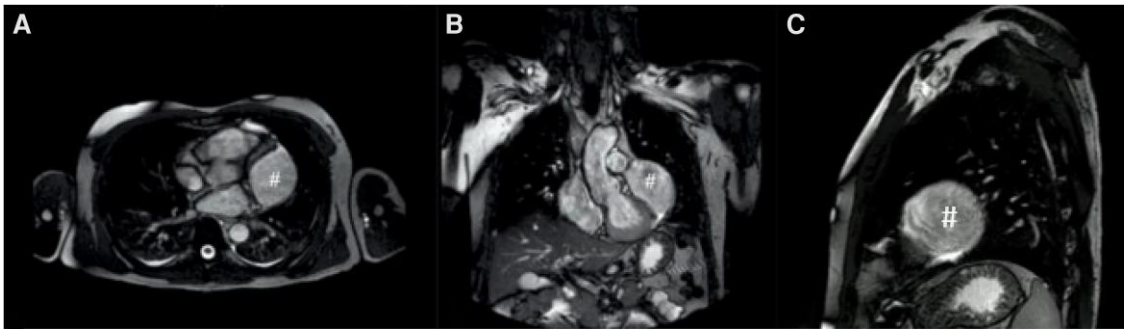


Figure 3 Cardiac magnetic resonance. (*A*) Transversal view. (*B*) Frontal view. (*C*) Sagittal view. # points the left atrial appendage aneurysm.

Table 1 Clinical characteristics of patients with left atrial appendage aneurysm according to published literature

Study	No.	Age	Sex	Dimensions	Palpitations	Dyspnoea	Chest pain	Cardio-embolism	Treatment					Prognosis	
									None	Thoracotomy	Mini-thoracotomy	AF ablation	Medical therapy	Death	Symptoms
Aryal *et al.* 2014[1]	82	31 ± 20	54% M	7 ± 3 × 6 ± 2	43%	22%	7%	11%	16%	83% surgical therapy		NA	NA	NA	NA
Fakhri *et al.* 2020[3]	14	<18	71% M	NA	42%	28%	NA	21%	7%	92% surgical therapy		NA	7%	7%	7%
Hosseini *et al.*, 2016[6]	132	NA	50% M	NA	54%	27%	0.7%	12%	0%	81% surgical therapy		NA	NA	1.5%	5%/3.8
Wang *et al.*, 2017[7]	101	30 ± 20	48% M	11 ± 5 × 7 ± 3	45%	29%	NA	6%	34%	74%	10%	NA	11%	NA	NA

formations were seen with normal velocity flow measured via pulsed-wave Doppler (see Supplementary material online, *Videos S2–S3*).

To conclude, the patient underwent a cardiac magnetic resonance (CMR) which confirmed the kinetic alterations localized at the anterior wall of the left ventricle, where the LAAA (68 × 70 × 54 mm) was found (*Figure 3*).

Differential diagnosis with other conditions like pericardial cyst or lipomas was achieved by the analysis of the morphology of the mass, its anatomical relationship with the left atrium (LA), and the absence of enhancing at CMR T1-weighted imaging. The findings were discussed with the patients. The risks and benefits of the possible therapeutic options were clarified, and he expressed the preference not to undergo invasive open-heart surgery. Therefore, the case was discussed with the heart team of our hospital, and, in agreement with the patient's will, strict follow-up was programmed. Moreover, due to the high risk of appendage perforation, transcatheter ablation for atrial fibrillation was deemed unsuitable. Finally, the patient was monitored using an implantable loop recorder.

Considering the history of mild arterial hypertension, we evaluated his thromboembolic risk to be higher than his bleeding risk; therefore, the patient started a direct oral anticoagulant (DOAC) (edoxaban) to prevent the formation of thrombi in the appendage and flecainide (50 mg bid) and metoprolol tartrate (25 mg bid) for rhythm control.

At a 1-year follow-up, the patient was asymptomatic with no further arrhythmic episodes found during the loop recorder interrogation.

Discussion

The first report of an aneurysmatic left atrial appendage was described in 1960.[1] To date, fewer than a hundred cases have been reported, with a mean presentation age of 30 ± 20 years and a higher prevalence among females,[2] making it very difficult to draw solid evidence from the available studies.

Left atrial appendage aneurysm have been classified in congenital and acquired, with the latter being more prevalent.[4]

Congenital forms seem to be secondary to a fibrotic evolution of the pectinate muscles and of atrial muscle fibres.[5] Left atrial appendage aneurysm can occur in association with other congenital anomalies such as atrial septum defect, ventricular septum defect, anomalous renal artery, and Noonan syndrome.[2] Acquired aneurysms are most commonly secondary to atrial enlargement due to mitral and/or inflammatory diseases.[3] The most common clinical presentations include palpitations and concomitant dyspnoea with subsequent finding of supraventricular arrhythmias such as atrial fibrillation or flutter. Thrombotic formations may be the first clinical manifestation[5] (*Table 1*).

Finally, chest pain has been described as a clinical manifestation of LAAA, possibly secondary to myocardial ischaemia due to external compression of the left anterior descending coronary artery.[7]

Compression may rarely impair ventricular filling, determining signs and symptoms like cardiac tamponade.[8]

It is frequent[8] to find a mass in the mediastinum which may cause deformation of the heart dimensions when assessed through chest X-ray. However, given the reduced specificity of this method, such a finding is not useful in the differential diagnosis with left ventricular wall aneurysms, pericardial cysts, or mediastinal masses.

Transthoracic echocardiogram has a low sensibility in diagnosing the aneurysm; it is however useful to assess ejection fraction and valvular defects that could have been caused by external compression.[9]

The diagnostic gold standard is represented by transoesophageal echocardiography which allows to evaluate the saccular morphology, the neck, the flow velocities of the LAA, and the presence of thrombi of congenital anomalies.[8] Cardiac magnetic resonance imaging allows an accurate assessment of the LAAA dimensions and of its anatomical relationships with other surrounding structures. It also analyses 4D blood flows that can help determine the thromboembolic risk.[10,11] If coronary compression is suspected, a coronary angiography may be useful.

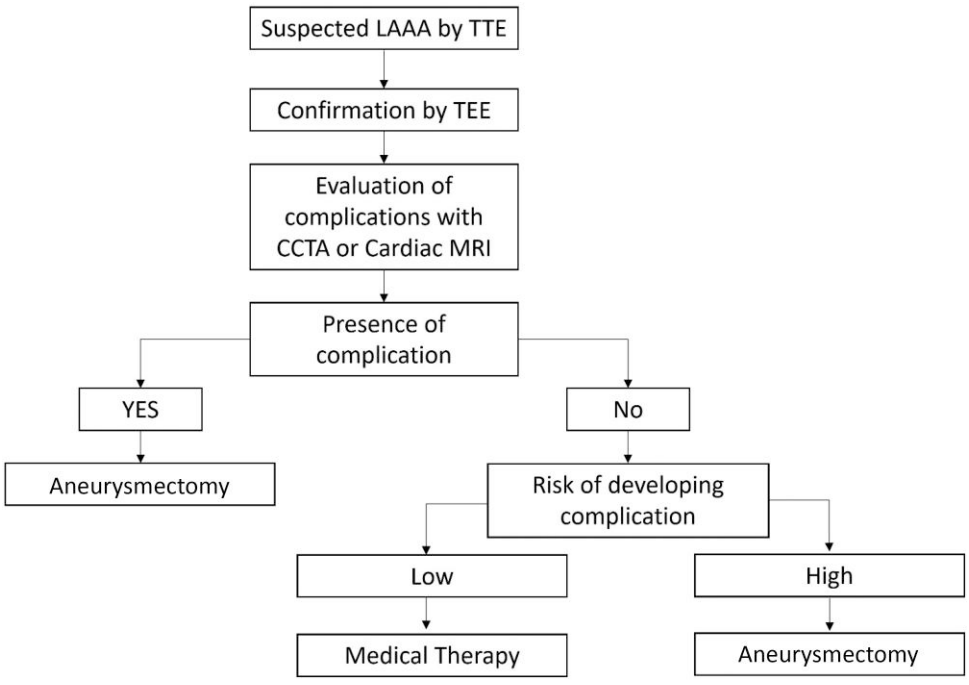


Figure 4 Proposal for a diagnostic and therapeutic algorithm for patients with left atrial appendage aneurysm.

Most of the case reports that can be found in the current literature suggest surgical treatment (aneurysmectomy) even if the patient is asymptomatic to avoid complications.[5] Median sternotomy is the most used approach; however, a minimal invasive thoracoscopy can be performed in the case of small aneurysms.[12] Cardiopulmonary bypass could be avoided if there are no thrombi in the LAAA.

It is known that atrial appendage could play a hormonal role in varying levels of insulin, leptin, and adiponectin, although the hormonal or haemodynamic consequences of surgical LAAA exclusion are not known.[6]Moreover, in the case of atrial fibrillation, a surgical ablation procedure (Cox Maze III) can be associated.[12]

Post-surgical prognosis is usually favourable at 180 days of follow-up.[11] A simplified diagnostic and therapeutic algorithm for patients with LAAA is proposed in *Figure 4*.

Asymptomatic patients who refuse surgical therapy can undergo a conservative approach if the LAAA dimensions are stable. Pharmacological therapy to reduce thromboembolic risk as well as anti-arrhythmic prophylaxis can be prescribed[8] (*Table 1*). Although there is no data to choose between DOACs or vitamin K antagonists (VKAs), we do not see a pathophysiological reason not to use a DOAC that remains the first choice in cases of concomitant atrial fibrillation. If there are no atrial arrhythmias, the role of anticoagulation is unknown.

This lack of information between different types of anticoagulants is due to the fact that most cases described in literature belong to the pre-DOAC era. Moreover, the CHA2Ds-VASc scoring system was not studied in these cases.

Because of the rarity of this condition and the lack of prospective studies that confront medical and surgical therapy, the treatment of this patients should still be based on a case-by-case scenario.

Conclusions

Left atrial appendage aneurysm is a rare clinical condition that can lead to a significant impairment in the patient's life, it has a heterogeneous clinical presentation, and the diagnosis is achievable through multimodality imaging. There is not enough evidence to have a general consensus on how to treat this condition; therefore, an accurate evaluation of every case should be done. Every case should be managed according to the single patients' needs, analysing symptoms (palpitations, dyspnoea, chest pain, syncope), dimensions of the aneurysm, relationships with other mediastinal structures (compression), the presence of endo-auricular thrombi, and finally, the patient's will. A multidisciplinary approach could be helpful in evaluating every single case.

Lead author biography

Francesca Coraducci is a resident in cardiovascular diseases at Marche Polytechnic University.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: Informed consent for publication was obtained from the patient. The authors confirm that consent for publication has been

obtained, in line with the Committee on Publication Ethics (COPE) best practice guidelines.

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Data availability

No new data were generated or analysed in support of this research.

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