A challenging management

Giant left atrial appendage aneurysm with thrombus

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Summary

We present the case of an atrial thrombus that developed under established anticoagulation in a patient with a giant left atrial appendage aneurysm and persistent atrial fibrillation. The management of patients with left atrial appendage aneurysm is challenging and might include surgical resection of the aneurysm and antithrombotic treatment tailored according to surgical risk, comorbidities and patient preferences.

Case description

An 86-year-old woman with a history of persistent atrial fibrillation anticoagulated with rivaroxaban (CHA2DS2-VASc score 4, HAS-BLED score 3) was referred to our emergency department because of suspected acute gastrointestinal bleeding with progressive anaemia requiring the transfusion of two units of packed red blood cells. Prompt endoscopic evaluation revealed no gastrointestinal source of bleeding and the anticoagulation was resumed after having been paused for a few days. The clinical course was complicated by a transfusion-associated circulatory overload requiring intravenous diuretics. Subsequent transthoracic echocardiography showed a preserved left-ventricular ejection fraction, mild to moderate mitral valve regurgitation and massively enlarged atria. Cardiac computed tomography confirmed the presence of an aneurysmatic enlarged left atrial appendage (80 × 46 × 25 mm, volume 99 ml) without signs of mechanical compression of the surrounding anatomical structures, and a floating thrombus (17 × 34 × 12 mm) adhering to the inferomedial wall of the left atrium.

In light of the size of the thrombus and the short interuption in anticoagulation we postulated that the thrombus had been present already before hospital admission. According to the patient's wish, advanced age and comorbidities, we proposed a conservative treatment. The oral anticoagulation was switched from rivaroxaban to phenprocoumon. The patient was discharged to a nursing home but suddenly died 4 weeks later of an unknown cause, likely of cardiac origin.

Discussion

Left atrial appendage (LAA) aneurysm is a rare condition that can be congenital or – more commonly – acquired. It is synonymously described as “giant LAA”, “third ventricle” or “fifth chamber”, usually when its length exceeds 5 cm. Little is known about the prevalence in the general population with only a few hundred cases reported in the medical literature. The aetiology of congenital forms is thought to be related to congenital dysplasia of the pectinate muscles and the left atrial muscle bundles, whereas acquired forms appear mostly in association with elevated left atrial pressure due to mitral valve disease or left ventricular dysfunction [1].

According to the largest systematic review, patients with an LAA aneurysm have a broad spectrum of age at diagnosis, ranging from the prenatal period to 88 years (mean 31 years), are frequently asymptomatic or might report palpitations (43%), dyspnoea (22%) and/or thromboembolic events (11%) [2]. Progressive dilation of the LAA aneurysm may compress the surrounding structures including the left recurrent laryngeal nerve resulting in hoarseness. This constellation is referred to as cardiovocal syndrome (Ortner syndrome). It was not the case with our patient.

While signs of left atrial enlargement can be shown on conventional chest X-ray, diagnosis is mostly established by transthoracic and/or transoesophageal echocardiography, which can also provide information about the presence of intracardiac thrombi. Cardiac computed tomography or magnetic resonance tomography may assist diagnosis and can further assess the compressive effect on the surrounding structures. The prevalence of thrombi in an LAA aneurysm is high (~25%) and likely due to blood stasis in the aneurysmatic cavity. However, the risk of cardioembolic events seems to be mostly associated with the occurrence of atrial arrhythmias and less with the size of the aneurysm [2]. The role of LAA aneurysms as a substrate for atrial fibrillation is controversial but some authors discuss the aneurysm as a focus of atrial arrhythmia. Therefore, pulmonary vein isolation might be an ineffective treatment option for a patient with atrial fibril-

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lation and LAA aneurysm, to maintain sinus rhythm [3].

Despite the lack of evidence-based guidelines for the management of LAA aneurysm, surgical resection is frequently recommended in patients at low surgical risk, particularly in symptomatic individuals and in those with evidence of thrombi, to prevent further potential life-threatening complications. Some authors recommend surgery even in asymptomatic patients at low surgical risk. Resection of the aneurysm is considered curative with generally excellent outcomes and very low procedural mortality. After surgical resection, in the absence of atrial fibrillation, anticoagulation may be discontinued. We strongly believe that, although this might be valid for congenital forms with isolated forms of LAA aneurysm, surgery alone is rarely curative in acquired forms associated with other cardiac pathologies. In these cases, discontinuation of anticoagulation in patients with concomitant severe enlargement of the left atrium and/or atrial arrhythmias should be discouraged.

Transcatheter LAA occlusion is also a therapeutic option to reduce the risk of thrombus formation, but frequently not feasible in cases of giant LAA aneurysm because of technical issues, in particular related to the absence of such large occluders. Double-device LAA closure might be a feasible alternative in selected cases.

When surgery is performed, a Cox-maze III or other ablative procedures are frequently performed to treat or prevent atrial fibrillation, particularly in patients with giant aneurysms or atrial dilation.

In addition, antithrombotic treatment to prevent cardioembolic events should be evaluated. In the absence of specific trials for the management of LAA aneurysm, the following elements should be considered in decision-making.

Firstly, the overall prevalence of thrombi and the occurrence of cardioembolic events are very high among patients with LAA aneurysms, in particular when some anatomical features are present. Therefore, it is our opinion that oral anticoagulation is likely to be beneficial in patients without atrial fibrillation in the absence of strong contraindications. However, studies are lacking because the condition is very rare.

Secondly, the presence of atrial arrhythmia is associated with an even higher risk of cardioembolic events. It is our opinion that all patients with LAA aneurysms and atrial fibrillation should receive an oral anticoagulant to prevent cardioembolic events. Of note, the use of common scores to estimate the embolic and bleeding risks (CHA2DS2-VASc score, HAS-BLED score) is not validated in this particular population. No specific data exist in favour of or against the use of direct (non-vitamin K antagonist) oral anticoagulants (NOACs) in this context. The occurrence of cardioembolic events does not seem to be correlated with anatomical features of the aneurysm; however a careful clinical and echocardiographic examination may reveal additional comorbidities (e.g., mitral stenosis) that might influence the decision about the optimal class of antithrombotic drug. Of note, thrombi might arise outside the LAA aneurysm, as observed in our patient. Special consideration should be given to the particular anatomy and the extreme blood stasis, the efficacy and safety profile of vitamin K antagonists and NOACs, and the exclusion criteria of trials investigating NOACs (i.e., patients with moderate/severe mitral stenosis and mechanical prosthetic valve) [4].

If an LAA thrombus arises or persists during follow-up despite confirmed good adherence to a NOAC regimen, an individualised management strategy is required.

Thirdly, in patients with LAA aneurysms and evidence of atrial thrombus the further medical management may be challenging, in particular, if thrombus appears despite the correct use of oral anticoagulants.

Generally, the standard therapy in patients with evidence of LAA thrombus consists of vitamin K antagonists with rigorous follow-up and international normalised ratio (INR) monitoring until resolution of the thrombus [4]. However, similar rates of thrombus resolution have been observed in several small recent studies investigating the NOACs rivaroxaban, apixaban, and dabigatran. Together, these data indicate that use of NOACs is an alternative option in the case of evidence of LAA thrombus, particularly when a vitamin K antagonist is not well tolerated or adequate INR control cannot be obtained. If an LAA thrombus arises or persists during follow-up despite confirmed good adherence to a NOAC regimen, an individualised management strategy is required [4]. This may include switching to a different type of NOAC (direct thrombin inhibitor to factor-Xa inhibitor or vice versa) or INR-tailored vitamin K antagonist therapy (mostly INR 2.5–3.5 until complete resolution of thrombus) [5].

No recommendations exist on the optimal treatment of thrombi in patients with LAA aneurysms. We performed a systematic search of the available literature in the PubMed and EMBASE databases, using medical subject heading (MESH) “LAAA”, “left atrial appendage aneurysm”, “left atrial aneurysm” and “thrombus” or “therapy”. As of October 2021, 18 case reports were found. Ten described a curative surgical resection

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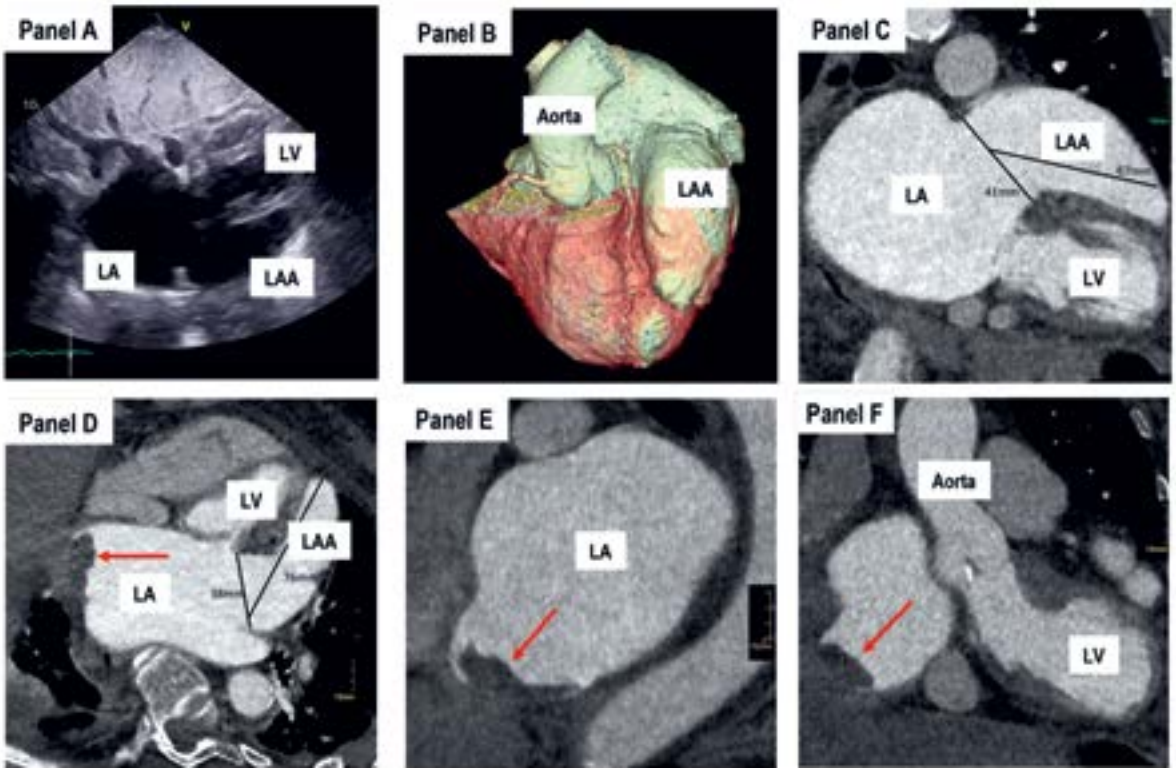

Figure 1: Panel A: Echocardiographic subcostal view showing the left ventricle (LV), the left atrium (LA) and the aneurysmatic left atrial appendage (LAA). Panel B: 3D reconstruction of the heart showing the LAA aneurysm. Panel C: Two-chamber view showing the size of the LAA aneurysm in comparison to the LV. Panels D to F: Modified axial, short axis and three chamber views showing the thrombus in the left atrium (red arrow).

of the aneurysm without further anticoagulation, eight (45%) described conservative strategies with antithrombotic drugs (three cases with useof a vitamin K antagonist, one case with dabigatran, and four cases without further specification). In four cases resolution of the thrombus was achieved, in one case the thrombus persisted despite anticoagulation with phenprocoumon and in three cases there was no follow-up described.

In our patient, a large atrial thrombus was observed despite treatment with rivaroxaban for atrial fibrillation. As such, we decided to switch to a vitamin K antagonist therapy. Follow-up, however, could not be performed because the patient died 4 weeks after discharge.

In summary, LAA aneurysm is a rare condition, frequently asymptomatic but associated with a high risk of cardioembolic events, in particular in patients with atrial fibrillation. The optimal treatment may include surgical resection of the aneurysm and antithrombotic treatment to reduce the risk of such events. In the absence of dedicated studies, the management should be tailored according to surgical risk, comorbidities, and patient preferences.

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