Congenital left ventricular aneurysm: a cause of impaired myocardial torsion and peripheral thrombo-embolic events

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Congenital aneurysms of the left ventricle (LV) are rare cardiac abnormalities and in most instances, are asymptomatic. However, some patients may present cardiac rupture, tamponade, ventricular arrhythmias, and eventually sudden death. Herein, we describe a case of a 64-year-old male patient who was hospitalized for critical limb ischaemia because of an acute embolic event. Transthoracic contrast echocardiography revealed a congenital aneurysm of the LV apex with a small thrombus and a reduced LV ejection fraction. Speckle tracking imaging showed an impaired myocardial torsion. Diagnosis was confirmed after surgical resection of the aneurysm. LV torsion and ejection fraction were normalized after surgery.

Keywords

Contrast • Aneurysm • Speckle tracking • Torsion • Thrombo-embolism

Introduction

Congenital left ventricular (LV) aneurysm is a rare cardiac abnormality with a prevalence of 0.34% in adult patients.¹ This type of aneurysm may be located in the atria,² on the left atrial appendage,³ but most frequently in the LV. Patients with congenital LV aneurysm usually remain asymptomatic and their aneurysm is incidentally diagnosed at a mean age of 43 years. However, some patients may present supraventricular or ventricular arrhythmias, tamponade, cardiac rupture, or even sudden cardiac death.⁴ LV diverticulum is another a rare cardiac anomaly with similar morphological characteristics to congenital LV aneurysm.⁴ Congenital LV aneurysm and diverticulum

Figure 1

Two-dimensional echocardiography (four-chamber view) revealed an aneurysm of the left ventricular apex (A). Echocardiography after contrast infusion (B) detected a mobile thrombus (arrow) into the aneurysm.
are a potential sources of emboli and transthoracic echocardiography (TTE) remains a standard tool for their differential diagnosis.

**Case presentation**

A 64-year-old male patient was admitted to our hospital because of a stabbing pain in his right lower extremity. The clinical examination revealed a pale and pulseless right leg indicating acute limb ischaemia. Electrocardiogram showed sinus rhythm with non-specific T-wave inversion in II, III, aVF, V3 to V6 leads. Laboratory tests showed an elevated creatine phosphokinase level (2225 mg/dL) whereas troponin levels were below normal values. The patient had a history of diabetes mellitus type II and no previous history of coronary artery disease or myocardial infarction. The patient underwent urgent embolectomy of the right femoral artery and a large thrombus (2 × 3 cm) was excised. TTE (Vivid 7, GE Medical System, Horten, Norway) revealed a cavity with a narrow neck located at the LV apex, showing a paradoxical motion (dyskinesia) and absence of systolic wall thickening which were indicative of an aneurysm (Figure 1A). Contrast echocardiography (continuous infusion of 1 mL/min Sonovue, Bracco Italy) also confirmed the presence of a mobile thrombus into the aneurysm, which was presumably the source of emboli in the right femoral artery (Figure 1B). The calculated LV ejection fraction after exclusion of the LV aneurysm area was 45%. Furthermore, we measured myocardial rotation at the mitral valve level and at the apex just before the neck of the aneurysm from the short-axis view. Using commercially available two-dimensional strain software (Echopac Software GE Medical System, Horten, Norway), the endocardial border of the end-systolic frame was manually traced. A region of interest was then drawn to fit the entire myocardial wall thickness. Peak torsion (degrees) (white line in Figure 2) was automatically calculated as the difference between the apical rotation (green line in Figure 2) and basal rotation at the mitral valve level (red line in Figure 2). Speckle tracking imaging revealed a reduced LV torsion (8.4°, Figure 2) mainly attributed to a reduced apical rotation.

Subsequent cardiac magnetic tomography confirmed that the LV aneurysm had a thin fibrotic wall without myocardial cells which was covered by pericardium (four-chamber view). Cardiac magnetic tomography confirmed that the left ventricular aneurysm had a thin fibrotic wall without myocardial cells which was covered by pericardium (four-chamber view).
A thrombus was found attached its wall, near the neck. Histopathologic evaluation showed fibrous tissue without myocardial fibers, which was compatible with a congenital LV aneurysm. The patient had an uneventful postoperative recovery. A subsequent, trans-thoracic echocardiogram with contrast showed an improvement in LV systolic function (ejection fraction $=60\%$) ($\text{Figure 4A and B}$) related with a parallel increase in peak torsion ($12.5^\circ$, $\text{Figure 5}$).

**Discussion**

The prevalence of congenital LV aneurysm is $\sim0.34\%$ in the adult population, though large epidemiological data are lacking. This type of aneurysm is commonly located at the LV apex and is asymptomatic. However, a patient may experience supraventricular or ventricular arrhythmias, tamponade, cardiac rupture, or even sudden cardiac death.$^4$

The differential diagnosis of congenital LV aneurysm includes LV diverticulum. Congenital LV aneurysm has a large LV communicating neck with a fibrous wall and a paradoxical motion (dyskinesia). In contrast, LV diverticulum, in most cases, has a narrow neck with a muscular wall and a synchronous contraction with LV.

Furthermore, congenital LV diverticulum is usually detected in children as part of Cantrell’s pentalogy. Conversely, congenital LV aneurysm is not related with other pathology.$^5$ In our case,
the dyskinetic and thin walls of the outpouching cavity and the absence of synchronous contraction with the LV were the major characteristics supporting the diagnosis of congenital LV aneurysm vs. LV diverticulum. Cardiac MRI may also assist the identification of the fibrotic walls and the lack myocardial fibres confirms the diagnosis of congenital LV aneurysm.\(^6\)

As far as the prognosis is concerned, congenital LV aneurysm is usually associated with an adverse outcome, while LV diverticulum has a benign prognosis.\(^5,7\) Thrombo-embolic events are a frequent complication in patients with congenital LV aneurysm.\(^1\) We have presented a case of a 64-year-old male patient who was hospitalized for critical limb ischaemia because of an embolus originating from a congenital LV aneurysm as diagnosed by two-dimensional and contrast echocardiography. The tissue characteristics of the apical aneurysm as assessed by MRI, the lack of a previous history of an acute coronary syndrome, the absence of significant coronary stenoses in coronary angiography also support the diagnosis of the congenital origin of the LV aneurysm in our case. Additionally, the change of LV geometry caused by the bulging LV aneurysm was related with an impairment in LV function and a low myocardial torsion. After surgical excision of the aneurysm, both LV ejection fraction and myocardial torsion were improved.

In conclusion, congenital LV aneurysm is a potential source of emboli and may be related with an impairment of the overall LV function and deterioration of myocardial torsion. TTE is a useful tool to diagnose this cardiac anomaly. In a patient with a thrombo-embolic event, congenital LV aneurysm or diverticulum should be taken into consideration.

**Conflict of interest:** none declared.

**References**