Case report

Giant left atrial appendage aneurysm or pericardial effusion: a misleading appearance on echocardiography and a high risk of rupture

ABSTRACT

Left atrial appendage (LAA) aneurysm or giant LAA is an uncommon condition. It could be discovered incidentally during echocardiography examination or in symptomatic patients presenting with tachycardia or embolic events. The giant LAA is a serious condition with a high embolic risk that can cause respiratory distress and even cardiac arrest in children. A conservative surgical approach based on resection of the LAA is recommended and is mostly safe. We report the case of a four-year-old girl presenting with a symptomatic giant LAA removed successfully. The echo-cardiographer must be aware of its appearance mimicking a pericardial effusion and focus on its possible association with other congenital lesions. There are several therapeutic strategies, all with proven efficacy and safety.

Keywords: Giant Left atrial appendage aneurysm, pericardial effusion, echocardiography

1. INTRODUCTION

Left atrial appendage aneurysm (LAAA) is an usual condition. LAAA may be congenital: related to the dysplasia of the cardiac musculature, or acquired: complicating a mitral valve disease [1]. Most cases are reported in adults and the LAAA in infant is quite rare [2]. In general, the discovery is incidental and the patients could be asymptomatic. However, in some cases symptoms occur, including dyspnea, chest pain, supraventricular tachycardia, embolic events, and heart failure. The diagnosis is based on imaging which is fundamental in particular transthoracic echocardiography (TTE), multi-modal imaging, cardiac magnetic resonance imaging (MRI), 3D reconstructions and thoracic computed tomography (CT) scan are of great help in determining the limits, the exact measurements and the anatomical relationships with the surrounding organs [3]. In this report, we describe the case of an infant presenting with a symptomatic LAAA initially mistaken for pericardial effusion.

2. CASE PRESENTATION

A four-year-old female non consanguineous infant with no past medical history, presenting dyspnea and chest pain one month before her admission. Chest X-ray was realized demonstrating an enlargement of cardio-thoracic index. She, then, underwent fast-echocardiography revealing a large pericardial effusion lateral to the left ventricle. She was, therefore, referred to our cardiology center. Physical examination found a heart rate at 105 beats per minute, a blood pressure of 87/42 mmHg, no fever, normal cardiac and lung auscultation. Her electrocardiogram (EKG) found a sinus rhythm.
She underwent a TTE (figure 1) during that initial visit which revealed:

1. severe left atrial enlargement (LA volume of 41 mL/m^2),
2. dysplasic mitral valve leaflets without significant regurgitation nor stenosis

large extrinsic structure of antero-lateral location hypoechoic infringing against the left ventricle (80x87x72 mm) with spontaneaous intra-structural contrast and without constituted thrombus (figure 1-2).

3. No hemodynamic repercussions or respiratory variations.

A subsequent chest X-ray showing an anterior overlying structure on the left ventricle (figure 2(A)). Also, cardiac CT angiogram identified a markedly enlarged LAA causing mass-effect on the basal anterior and lateral walls of the left ventricle (figure 2 (B+C)).
The patient was referred for a cardiothoracic surgery evaluation. We suggested a conservative treatment strategy given the risk of embolic events, rupture strangulation and compression and adiastolia.
Therefore, surgery was recommended. The patient underwent a median sternotomy and LAA resection with cardiopulmonary bypass. The LAA was large and was excised just above the attachment to the left atrium. No defect, nor pericardium effusion were identified indicating that the mass represented an intrapericardial LAAA. Her post-operative course was uncomplicated, the patient is still in follow up, with risk of developing supra-ventricular tachycardia.

3. DISCUSSION

Giant LAA or aneurysms are unusual, especially in pediatric population. Mostly discovered at the second and third decade of life, and occurs more frequently in females [4]. Giant LAA could be acquired or congenital.
The genesis of congenital giant LAA is mainly due to a dysplasia of the pectineal muscle at the entrance of the atrium. Moreover, the acquired giant LAA is mainly related to conditions elevating the atrial pressure in the left atrium particularly the mitral valve disease and myocarditis generating a weakening of the atrial wall [5].
LAA aneurysms can present with catastrophic events such as a stroke or other systemic emboli. However, in most cases the giant LAA is discovered incidentally. Symptoms mostly found are palpitations related to supraventricular tachycardias, chest pain, shortness of breath, and rarely embolic events that are considered a severe complication. Moreover, the patients might present symptoms of heart failure [6].
The first line diagnostic tool is the TTE, specifically the subxiphoid view. Trans-esophageal echocardiogram, cardiac CT scan, or cardiac MRI, allow the diagnosis, the search for complications and allow to identify the anatomical relations to the surrounding organs. [5].
Congenital aneurysms are typically associated to an enlargement of the left atrium. Two types are described: intrapericardial giant LAA or extrapericardial giant LAA. The extrapericardial is associated to a pericardial defect with a herniated portion of the atrial appendage and atrial wall [7].
In our case, the absence of significant mitral valve disease or other cardiac pathology is probably related to a congenital giant LAA in its intrapericardial form.
The general recommendation for the management of LAAA is surgical excision. The three commonly described aneurysmectomy approaches are via a midline sternotomy, left thoracotomy, and mini-thoracotomy [8].
Given the aneurysm's size, the presence of other cardiac defects or thrombus, the heart team decides on the therapeutic strategy.
Surgical excision with median sternotomy and cardiopulmonary bypass, aortic cross-clamping, and surgical resection of the aneurysm with left atrial reconstruction is the mainly used approach. Other less invasive techniques could be suggested in smaller giant LAA [9].
In our case, the decision of the surgical approach was based on the mechanical complications arising from the LAA size and risk of strangulation, hemodynamic complications such as impairing the relaxation/filling of the left ventricle, and the significant risk of systemic embolization.

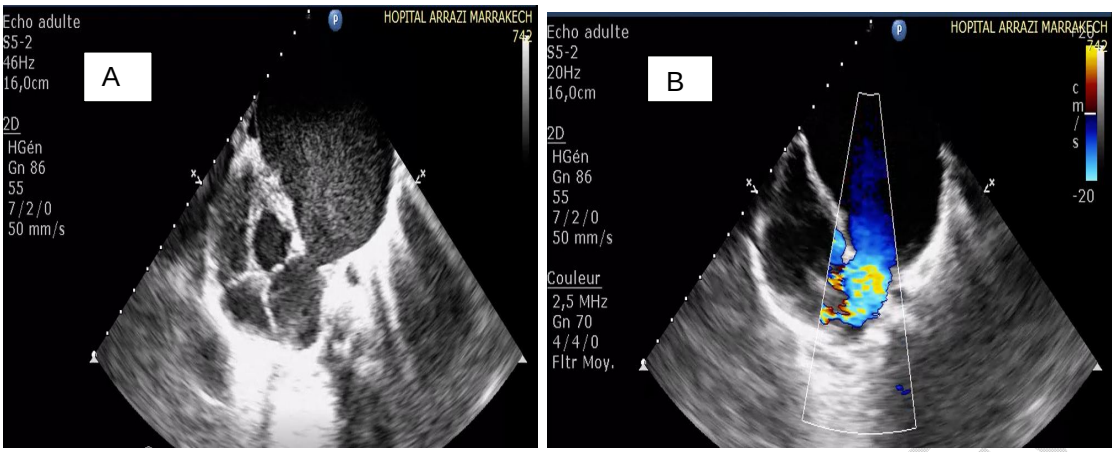

Fig. 1. Four chamber view showing large extrinsic structure of antero-lateral location hypoechoic infringing against the left ventricle with spontaneous intra-structural contrast (A). The cystic structure is in communication with the left atrium suggestive of a giant LAA (B).

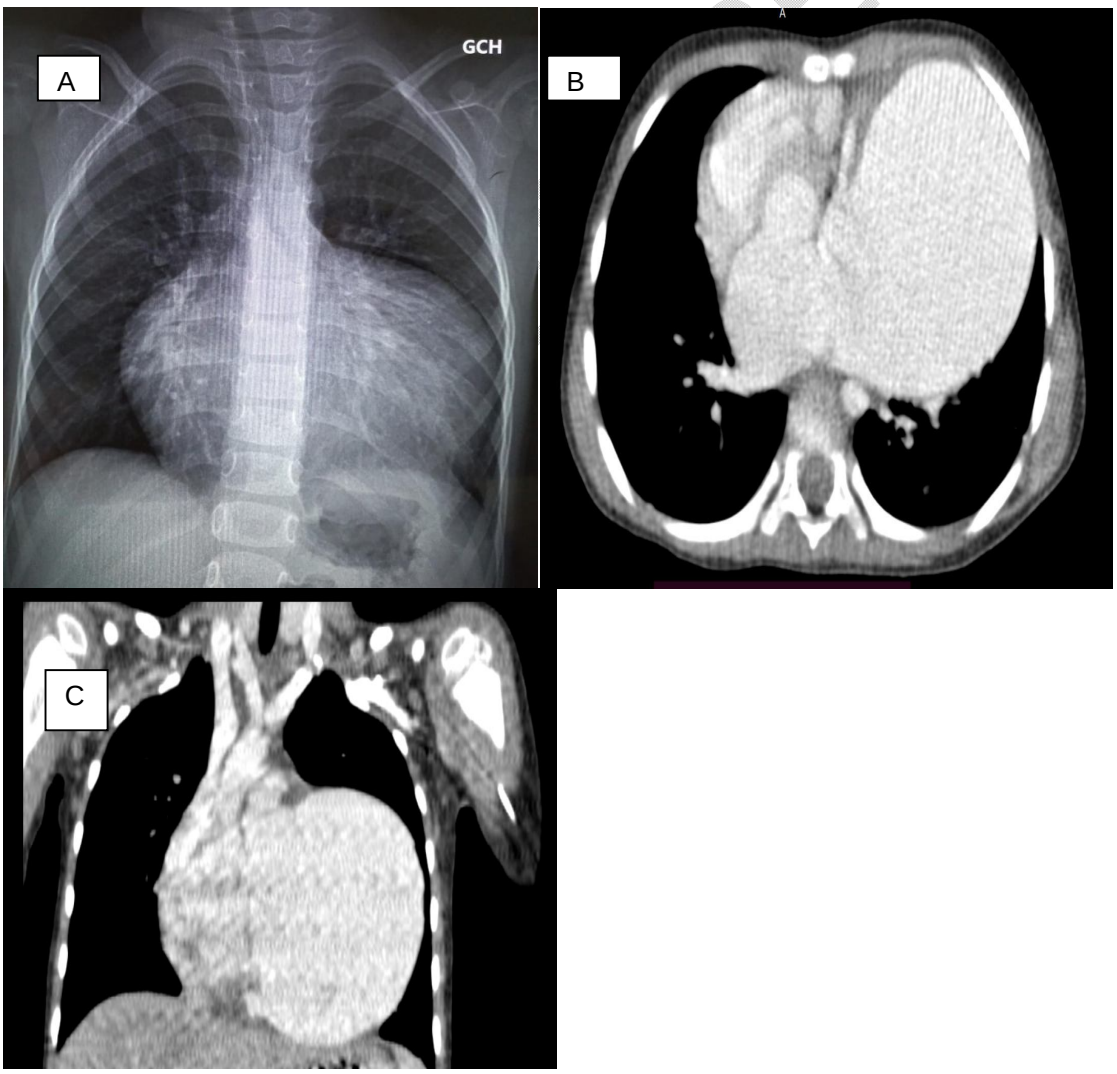

Fig. 2. Cardiomegaly with a structure superimposed on the left ventricle (A). Chest CT scan showing a giant LAA measuring 80 x 87 x 72 mm(B)+(C).

4. CONCLUSION

Our case demonstrates, that at an early age, the giant LAA might be symptomatic, and that the main differential diagnosis is pericardial effusion. The therapeutic decision must be made carefully, taking into consideration the clinical and echocardiographic parameters to a better management.

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