CASE REPORT

Idiopathic giant right atrial aneurysm

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ABSTRACT

A 2-year-old boy with an incidental finding of massive cardiomegaly on a chest X-ray was diagnosed with a giant right atrial aneurysm upon further investigation with echocardiography. The patient underwent successful surgical reduction of the right atrium and closure of the patent foramen ovale to prevent thromboembolic complications and to lower the risk of atrial arrhythmias. The resected atrium had paper-thin walls and pathological features of interstitial fibrosis with endocardial thickening.

Keywords: Cardiac magnetic resonance imaging, echocardiography, giant right atrial aneurysm

INTRODUCTION

Giant right atrial aneurysm is an extremely rare congenital cardiac condition that can present anywhere between fetal life and old age. Many cases are asymptomatic, but some present with atrial arrhythmias or thromboembolic complications. We report a case of an asymptomatic 2-year-old child who was incidentally diagnosed with a giant right atrial aneurysm and underwent successful surgical resection of the aneurysm.

CASE REPORT

A 2-year-old boy was brought to the pediatrician in view of non-specific respiratory symptoms where mother thought that he had a brief episode of apnea during sleep. The child was otherwise completely asymptomatic. He was then referred for a chest X-ray [Figure 1a] that showed significant cardiomegaly and therefore an echocardiogram was obtained. This showed a giant right atrial aneurysm (7 × 6 cm) [Figure 2a]. The tricuspid valve was anatomically normal with no stenosis or insufficiency. A small left-to-right shunt across a patent foramen ovale was also noted. Rest of the cardiac anatomy was normal. These findings were later confirmed by a cardiac magnetic resonance imaging (MRI) [Figures 3a and b]. Slow-flow artifacts were seen within the lumen of the right atrium without intracavitary thrombus. To our surprise, the electrocardiogram was normal. It showed normal sinus rhythm with normal P wave morphology and no rhythm abnormalities.

We opted to proceed with surgical resection of the aneurysm after discussions with the family, because of the risk of future thromboembolic complications and arrhythmias. Surgery was performed through median sternotomy. The pericardium was normal. A giant thin-walled aneurysm of the entire free wall of right atrium was noted [Figure 4a]. After establishing cardiopulmonary bypass, the aneurysm was opened. There was no evidence of thrombus or inflammation within the right atrial aneurysm. The aneurysm was resected as two pieces measuring 2 × 2 cm and 7 × 3.5 cm respectively. After excision of the affected right atrial wall, the defect was closed and care was taken not to compromise the right coronary artery.

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Direct suture closure of the patent foramen ovale was also performed. Histopathological examination of the resected atrial tissue showed paper-thin walls [Figure 4b], and the pathological features included interstitial fibrosis and endocardial thickening with no evidence of thrombosis.

The postoperative course was significant for short non-sustained runs of ectopic atrial tachycardia that were controlled on Atenolol. He was discharged home 4 days after surgery. The chest X-ray [Figure 1b] performed following surgery showed significant reduction in cardiac size. An echocardiogram [Figure 2b] performed prior to discharge showed significant reduction in the right atrial size, trivial tricuspid regurgitation, and a small pericardial effusion. At the time of first post-operative visit to the clinic a week from discharge, the effusion had resolved and there were no new findings on the echocardiogram.

**DISCUSSION**

Giant right atrial aneurysm is an extremely rare condition of unclear etiology. Some speculate an intrinsic structural protein defect, abnormal collagen or dysplastic pectinate muscles make the right atrium prone to dilatation even under low right atrial pressure.[1] There are few case reports in the literature to date since Bailey first reported this condition in 1955.[2] Majority of the patients are asymptomatic at presentation; some present with arrhythmias, palpitations, chest pain, shortness of breath or fatigue.[3] Age of presentation is widely variable from fetus to adulthood.[4] Many of the asymptomatic patients are identified by the presence of cardiomegaly on a chest X-ray, such as our case. Based on the X-ray alone other conditions such as Ebstein anomaly, cardiac tumors and pericardial cysts need to be considered. The diagnosis can be confirmed on echocardiogram and/or MRI as was done in our case. The tricuspid valve was thoroughly examined for any displacement or stenosis. While many cases with such a large aneurysm of the right atrium will have tricuspid annular dilation and resultant significant regurgitation of the tricuspid valve, this was not found in our case. Although sinus rhythm is seen in majority of the patients, such as our patient, various atrial arrhythmias can occur in about a quarter of the cases.[5] These include atrial flutter, atrial fibrillation, and supraventricular tachycardia. Patients are at an increased risk for intratral thrombus formation not only due to sluggish flow within the giant right atrium and atrial arrhythmias. This further predisposes them to thromboembolic complications including paradoxical and pulmonary embolism.[6] Whether surgery should be offered to asymptomatic patients remains controversial.[1,7] Given the high incidence of complications such as arrhythmias and thromboembolism, we recommend surgery even if the patients are asymptomatic.

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