Giant aneurysm of the atrial appendages in infants

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ABSTRACT

This case series describe two patients with giant aneurysms of the atrial appendages. This report discusses the clinical symptoms, imaging modalities, indications for surgical intervention, and histology of atrial appendage aneurysms.

Keywords: Atrium, congenital, dilation, pectinate muscles

INTRODUCTION

Giant aneurysm of an atrial appendage is a rare anomaly of unknown etiology although dysplasia of atrial appendage musculature has been proposed.[1,2] The right atrial appendage appears to be involved more often than the left atrial appendage. Most atrial appendage aneurysms are discovered fortuitously in asymptomatic adults.[1,2] However, a few have been detected in the fetus or in young infants, indicating that at least some aneurysms are congenital.[3,4]

We describe two infants who presented with an aneurysm of an atrial appendage documented by echocardiography and cardiac magnetic resonance imaging (MRI). Both presented with respiratory symptoms and poor growth and underwent resection of the aneurysm.

CASE REPORTS

Case 1

A female infant was born by uncomplicated spontaneous vaginal delivery at 40-weeks gestation following prenatal diagnosis of an aneurysm of the right atrium (RA) by fetal echocardiogram. The birth weight was at the 20th percentile. A chest X-ray demonstrated marked cardiac enlargement nearly obscuring the lungs [Figure 1]. A postnatal echocardiogram confirmed the fetal diagnosis and demonstrated a massively dilated RA, a patent foramen ovale, and small arterial duct [Figure 2]. The heart was otherwise normal. An electrocardiogram demonstrated a low right atrial rhythm but p-waves of normal voltage and no arrhythmia. The patient remained asymptomatic and was discharged at age 5 days receiving daily aspirin.

Serial cardiac MRI exams, performed at 3 days, 9 months, and 15 months of age, showed progressive dilation of the RA (indexed right atrial volumes 153 ml/m^2, 280 ml/m^2, and 383 ml/m^2, respectively). The remainder of the cardiac anatomy was normal.

At around 18 months of age, she began experiencing dyspnea, reduced exercise tolerance, and became unable to sleep in the supine position. Weight gain had stalled over the preceding 6 weeks, and she declined to the 10th percentile. She developed a prominent S4 on cardiac exam. A repeat cardiac MRI demonstrated stable right atrial enlargement [Figure 3].

At age 20 months, the patient underwent surgical resection of the aneurysm and closure of the foramen ovale. The walls of the giant right atrial aneurysm were noted to be nearly transparent [Figure 4]. She recovered uneventfully except for a few brief self-limited episodes of junctional tachycardia early postoperatively. Two months following surgery, she was asymptomatic with improved weight gain (35th percentile). A follow-up echocardiogram showed significant reduction of right atrial size.

Case 2

A 5-month-old girl presented with a several weeks history of mild respiratory distress and wheezing.

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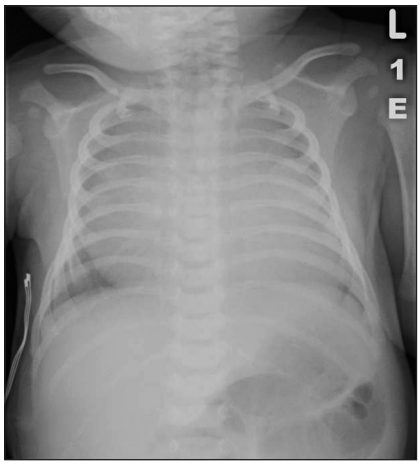

Figure 1: Frontal chest X-ray in case 1 demonstrating marked cardiomegaly

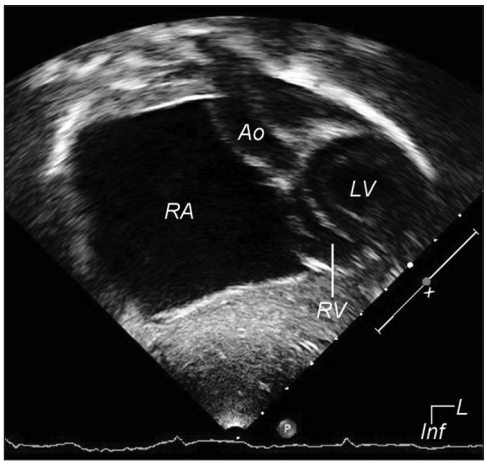

Figure 2: Subxiphoid sagittal view in case 1 showing a massively enlarged RA in comparison with the RV, LV, and Ao. RA: Right atrium, RV: Right ventricle, LV: Left ventricle, Ao: Aorta

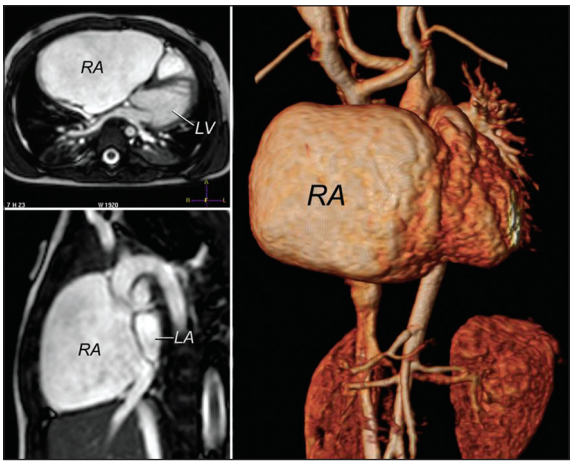

Figure 3: Cardiac MRI bright blood axial (upper left), bright blood parasagittal (lower left), and 3-D reconstruction (right) in case 1 showing the RA enlargement in comparison with the LA and LV. MRI: Magnetic resonance imaging, 3-D: Three-dimensional, RA: right atrium, LA: Left atrium, LV: Left ventricle

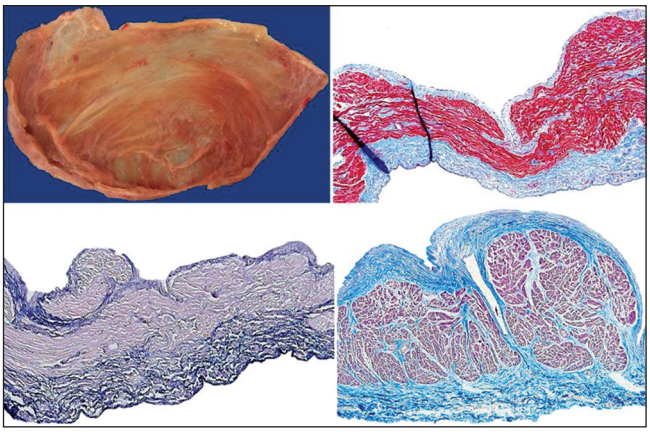

Figure 4: Upper left: Surgical specimen from case 1 showing the endocardial surface with few dispersed pectinate muscles and focal thinning of the wall. Upper right: Masson's trichrome stain of aneurysm showing abundant fibrous tissue (blue) and areas with thinning of muscle fibers (red). The endocardium appeared thinner than control. Lower left: Miller's elastic stain showing an abundance of elastin fibers especially in the epicardium. Lower right: Masson's trichrome stain of a section of RA in an age-matched control heart illustrating normal architecture. All sections are shown with endocardium superior and epicardium inferior. RA: Right atrium

A chest X-ray revealed marked enlargement of the cardiac silhouette and left lower lobe atelectasis. An echocardiogram showed a structurally normal heart with a large aneurysm connected by a broad neck to the left atrium (LA) between the left pulmonary veins and the atrioventricular junction [Figure 5]. Despite marked spontaneous cavitation, there was no thrombus. Mild hypoplasia of the left pulmonary artery was noted and flow in the lower lobe artery was bidirectional. There were no arrhythmias on surveillance monitoring. She underwent surgical resection of the aneurysm because of persistent left lower lobe atelectasis and respiratory symptoms. Surgical observation disclosed a thin-walled aneurysm of the left atrial appendage with pectinate muscles covering the internal surface and a small appendage tip at the lateral extent of the aneurysm [Figure 6]. The aneurysm was within an intact pericardium. The pulmonary veins were readily identified entering the thicker-walled portion of the LA and were not involved by the aneurysm. She recovered rapidly and a postoperative echocardiogram showed no residual aneurysm.

Pathology

Histologic sections of both aneurysms were compared with atrial appendage sections from age-matched controls that died from noncardiac causes and had a normal heart at autopsy [Figures 4 and 6]. Sections were stained with hematoxylin and eosin (H and E), Masson's trichrome, and Miller's Elastic. The specimen from case 1 was membranous tissue measuring 9.2 × 7.0 × 0.2 cm,

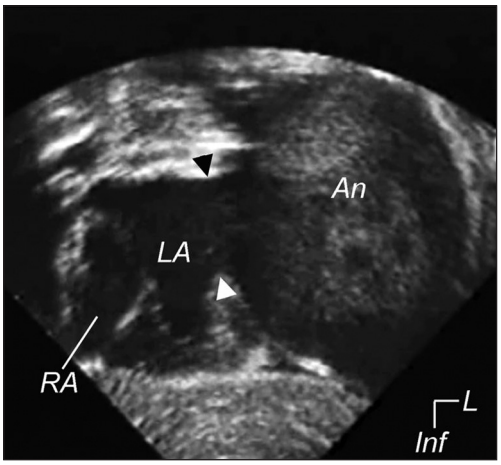

Figure 5: Subxiphoid long-axis view in case 2 showing a giant An of the left atrial appendage with spontaneous cavitation. The broad junction (arrow heads) between the aneurysm and the LA is seen. An: Aneurysm, LA: Left atrium

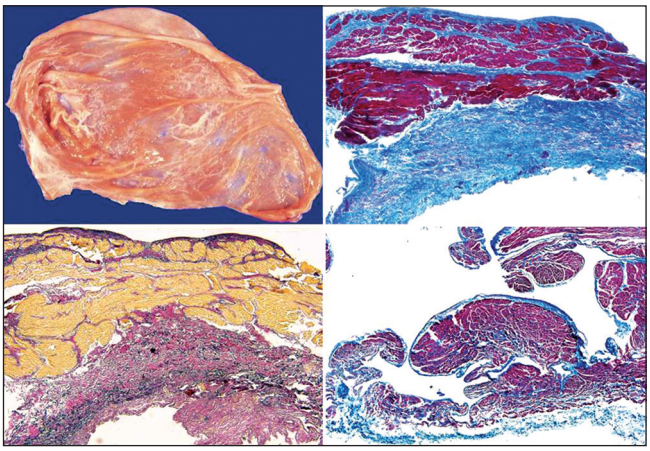

Figure 6: Upper left: Surgical specimen in case 2 showing dispersed pectinate muscles and relatively normal wall musculature interspersed with thin mostly fibrous segments. Upper right: Masson's trichrome stain of the aneurysm showing variable musculature (red) and increased fibrous tissue (blue), especially in the epicardium. Lower left: Miller's elastic stain showing marked increase in elastin fibers in the epicardium as well as between muscle bundles. Lower right: Left atrial wall in age-matched control heart illustrating the normal appearance. All sections are shown with the endocardium superior and the epicardium inferior

ranging from <1-2 mm in thickness. The epicardial surface was pink-tan and smooth and the transparent endocardium overlay brown musculature with a few dispersed pectinate muscle bundles. Focally, the wall was thin and devoid of muscle. Histologic sections showed a thickened wall. The endocardium and myocardium appeared thin and the epicardium thicker compared with the control.

The resected left atrial appendage aneurysm from case 2 measured 9.5 × 7.5 cm. The thickness at the base was 2-3 mm with some thin areas measuring <1 mm. The endocardial surface was smooth and transparent, and the epicardium was smooth and pink-tan. The pectinate muscles were remarkably deficient and focally absent in some areas. Histologic sections showed hypertrophied muscle bundles, a variable but mostly thin endocardium, and a thickened epicardium.

Both specimens showed an increase in coarse and thin elastic fibers in the epicardium, perivascular connective tissue, and between the cardiac muscle bundles. The endocardial elastic tissue was varied but not distinctly dissimilar from the controls.

DISCUSSION

These cases confirm that some atrial appendageal aneurysms are congenital and, as previously proposed, might be due to a developmental abnormality of the atrial musculature. Most atrial appendage aneurysms are discovered later in life, either fortuitously or because of an arrhythmia or embolic event.[1,2] Respiratory symptoms are an unusual manifestation in infants and children, especially for right atrial aneurysms.[5] Development of respiratory symptoms in case 1 corresponded to increasing size of the right atrial aneurysm. Divekar *et al.*, described a similar case of an enlarging right atrial aneurysm associated with episodes of respiratory distress mimicking severe bronchospasm.[6] In this case, dynamic distal airway collapse was demonstrated by fluoroscopy, probably due to increased intrapleural pressure generated to overcome the proximal obstruction. The clinical and fluoroscopic findings resolved following surgical repair. Our case 1 and that of Divekar *et al.*, highlight the potential for rapid enlargement of an atrial aneurysm in a young child. Case 2, discovered because of respiratory symptoms, underwent cardiac imaging for the first time at age 5 months so that the evolution of the left atrial aneurysm is not known. Neither of our cases had an arrhythmia or thrombosis — the more frequent manifestations of atrial aneurysm reported in older patients.

In both of our cases, histologic examination showed marked thinning of the wall of the resected aneurysm with reduced thickness of the atrial muscles and increased fibrous tissue and elastin fibers, especially in the epicardium.There were a few areas of complete loss of musculature, as reported previously in some cases.[7] We cannot determine if the appearance of the wall of the aneurysm was a primary developmental abnormality or caused by progressive dilatation of the aneurysm with stretching and thinning of the wall.

Charles Bailey first described congenital enlargement of the RA in 1955.[8] Binder *et al.*, reviewed 105 cases, almost all adults, of right atrial or coronary sinus aneurysms or diverticula reported between 1955 and

1998.[1] In this series, 60 patients had congenitally enlarged right atria, 28 patients had diverticulum of the coronary sinus, and the remaining patients had single or multiple diverticula of the RA. Of those with congenitally enlarged right atria, most (48%) were asymptomatic, but 28% complained of shortness of breath and 17% of palpitations. Less frequent symptoms included fatigue, chest discomfort, and syncope. A total of 28% of subjects in this review underwent intervention. In contrast, of those with diverticulum of the coronary sinus only 7% were asymptomatic, with the most common complaints being arrhythmias (46%) and palpitations (25%).[1]

Blaysat *et al.*, presented 15 pediatric cases gathered from several French centers.[5] A total of eight remained asymptomatic during follow-up, four developed an atrial arrhythmia successfully treated medically, and three underwent surgical resection because of a thrombus (2) or the large size of the aneurysm (1). All were alive and asymptomatic after 2-15 years (mean 6 years) follow-up.[5] While this is encouraging, the atriotomy scar has been implicated in producing atrial arrhythmias late after surgical resection.[6,9]

Forbes *et al.*, reviewed the indications for surgical intervention in children with an aneurysm of the RA, including atrial arrhythmias, thrombosis, progressive dilatation, airway compression, and tricuspid valve regurgitation.[9]

Left atrial appendage aneurysm is more rare. Chowdhury *et al.*, reviewed 78 cases reported between 1922 and 2007.[2] Most cases were detected in the third or fourth decade or later. An atrial arrhythmia was the initial manifestation in 60% and an embolic event in 18%. Other symptoms included dyspnea, ventricular dysfunction, mitral regurgitation from distortion of the valve annulus, and chest pain apparently from coronary artery compression. All but four of the patients in this review underwent resection of the aneurysm, which appears sufficient to abolish symptoms and embolic risk.[2]

Only a handful of cases of left atrial appendage aneurysms have been reported in children.[4] Most were asymptomatic but some presented with arrhythmia, stroke, or tamponade immediately after birth.[7] Resection of a left atrial aneurysm is almost always recommended because it effectively eliminates embolic risk and recurrence is exceedingly rare.[7]

Noninvasive imaging has facilitated the diagnosis and management of atrial appendage aneurysms. Echocardiography is the diagnostic modality of choice in infants and children and can exclude other causes of atrial enlargement.[10] Cardiac MRI or computed tomography can be used to confirm the diagnosis if necessary and exclude a pericardial cyst or mediastinal tumor, which might be confused with an atrial appendage aneurysm. Our case 1 demonstrates the utility of MRI for tracking enlargement of an atrial aneurysm, providing a better estimate of atrial size than echocardiography.[11]

Atrial appendage aneurysm is a congenital anomaly in some cases and can progress rapidly in young children. Respiratory symptoms are an unusual but important manifestation in infants and children. Echocardiography and cardiac MRI are effective for diagnosis and follow-up. Surgical intervention relieves symptoms, but long-term outcome has not been documented.

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