International Journal of Pediatrics and Adolescent Medicine (2016) xx, 1–3

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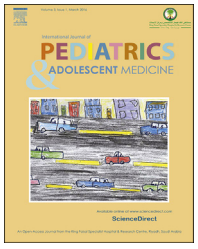

Case Report

Congenital aneurysm of the right atrial appendage

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Received 3 May 2016; received in revised form 13 July 2016; accepted 1 August 2016

KEYWORDS
Aneurysm;
Appendage;
Congenital;
Right atrium

Abstract Congenital aneurysm of the right atrial appendage is a rare cardiac anomaly with only a few reported cases in the literature. Most of the cases involved adults in their third decade of life. We report a case of congenital aneurysm of the right atrial appendage in a newborn, who initially presented with jaundice and an incidentally discovered systolic murmur. The diagnosis was established by an enhanced CT scan of the chest and an echocardiography that also showed an ASD and multiple VSDs. Because of its rare occurrence, diagnosis is difficult, and the symptoms may be confused with other causes of right atrial dilation, such as Ebstein's anomaly.

1. Introduction

Aneurysm of the right atrial appendage is a rare cardiac anomaly that most commonly occurs in adults. It is much rarer in the pediatric population, with less than 10 reported cases in the literature [1]. It can be identified during the prenatal period or incidentally thereafter during routine neonatal clinical examinations and can be associated with other complex cardiac anomalies [2–4]. It is important to distinguish this diagnosis from Ebstein's anomaly, which is a more common cause of right atrial enlargement [5–7].

2. Case report

A 1-day-old female infant was admitted to the pediatric ward as a case of neonatal jaundice to receive phototherapy treatment. The baby was born by normal spontaneous

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Peer review under responsibility of King Faisal Specialist Hospital & Research Centre (General Organization), Saudi Arabia.

http://dx.doi.org/10.1016/j.ijpam.2016.08.006

vaginal delivery at term, with a birth weight of 3.74 kg (just above the 50th percentile). There were no natal complications. On physical examination, the patient was jaundiced but otherwise was in good general health. Cardiac auscultation revealed a systolic murmur. The chest x ray showed an enlarged cardiac shadow with a prominent right atrial contour (Fig. 1).

A transthoracic echocardiogram was performed and showed a massively dilated RA, a moderately sized ostium secundum atrial septal defect, two muscular ventricular septal defects and a small patent ductus arteriosus with evidence of left to right shunt. The atrioventricular valves, the left atrium and both ventricles were normal with good ventricular systolic function (Fig. 2). An enhanced CT scan of the chest was requested and showed a large aneurysmal dilatation of the right atrial appendage with no internal thrombi (Fig. 3).

The patient remained stable and was discharged at 4 days of age on oral anticoagulation medicine (10 mg of aspirin daily), with an appointment at a cardiology clinic. At the first follow-up, the infant was asymptomatic with no significant changes in the echocardiogram. The plan was to follow up with the patient closely to monitor for the possible need for reparative surgery in the future.

3. Discussion

Aneurysms of the right atrial appendage are rare cardiac anomalies that most commonly affect adults in their third decade of life [1]. However, they can be seen in children and neonates and can also be detected prenatally, confirming the congenital nature of these malformations [2]. They can present with palpitation and dyspnea especially in older patients and can be, as in our case, completely asymptomatic [1].

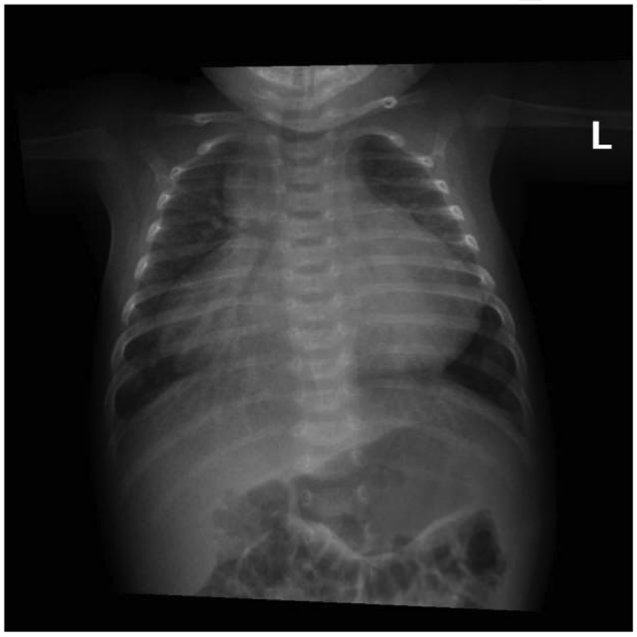

Figure 1 Chest X-ray, anteroposterior projection showing cardiomegaly with enlarged right atrial contour.

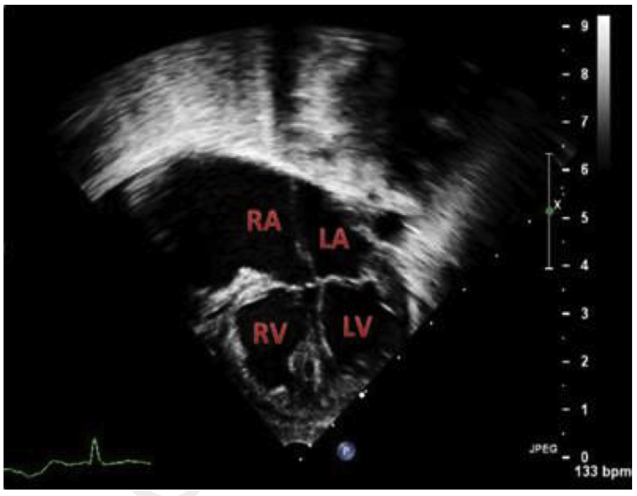

Figure 2 Transthoracic echocardiogram. The four-chamber view shows a massively dilated RA with normal position of the atrioventricular valves. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Echocardiography is the imaging modality of choice for diagnosis and follow-up because it is non-invasive, has no risk of radiation, can detect other congenital heart abnormalities and can evaluate other causes of right atrial enlargement [1,4,8,9]. Cardiac CT scans can also be performed to confirm the diagnosis and to identify any associated vascular abnormalities [9]. Multiple associated congenital cardiac anomalies have been reported, such as atrial and ventricular septal defects with shunting [10]. The top differential diagnosis of right atrial enlargement in these cases is Ebstein's anomaly, which can be easily recognized by its characteristic displacement of the tricuspid valve toward the right ventricle [11].

The management of right atrial appendage aneurysms is a matter of debate because the long-term outcomes of conservative versus surgical treatments have not yet been studied. In some studies, surgical treatment was effective

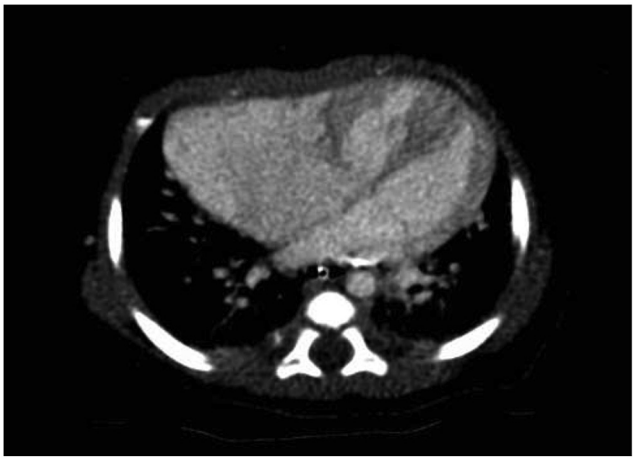

Figure 3 Enhanced axial CT scan of the chest showing a large dilatation of the right atrium and atrial appendage with a thin atrial wall.

Please cite this article in press as: Faqeeh S, et al., Congenital aneurysm of the right atrial appendage, International Journal of Pediatrics and Adolescent Medicine (2016), http://dx.doi.org/10.1016/j.ijpam.2016.08.006

Table 1 Reported cases of right atrial appendage aneurysms in infants.

Outcome	Treatment	Age and presentation	
Stable after surgery	Surgical correction	Infant with ectopic atrial tachycardia	Mizui et al., 2001
Stable at 1-year, 6-months follow-up	Medical treatment	Prenatal diagnosis	Tejero-Hernández et al., 2012
Stable at 8-months follow-up	Careful observation	Prenatal diagnosis	Ishii et al., 2012
Stable at 2-months follow-up	Surgical correction	Prenatal diagnosis	Lang et al., 2014
Stable at 4-months follow-up	Surgical correction	Prenatal diagnosis	Tunks et al., 2015
Gradual regression throughout the follow-up	Medical treatment	Prenatal diagnosis	Cardiel Valiente et al., 2016 (2 cases)
Stable throughout the follow-up	Careful observation	Prenatal diagnosis	

in preventing thromboembolisms and lowering the risk of atrial arrhythmia, which is one of the most common complications of these aneurysms [3,7]. In other studies, especially those that reported neonatal cases, conservative treatment in the form of oral anticoagulant medications was carried out to reduce the risk of thromboembolisms [10]. (Table 1).

4. Conclusion

Right atrial appendage aneurysms can be diagnosed in asymptomatic newborns, and treatment should be modified according to the age, presentation, other imaging findings and the follow-up results for more effective treatment plans.

Conflict of interest

None declared.

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