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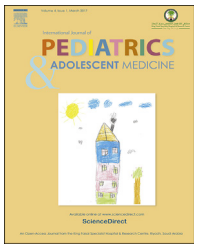

Instructive Case

Congenital aneurysm of the right atrial appendage

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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 incidentally discovered systolic murmur. The diagnosis was established by enhanced CT scan of the chest and echocardiography that also showed atrial septal defect (ASD) and multiple ventricular septal defects (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 diagnosed during adulthood. It is much rarer in the pediatric population, with less than 10 reported cases in the literature [1]. It can be identified during 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 in this age group [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 vaginal

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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 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 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 therapy (10 mg of aspirin daily) with an appointment for the 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 for the possible need of 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].

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

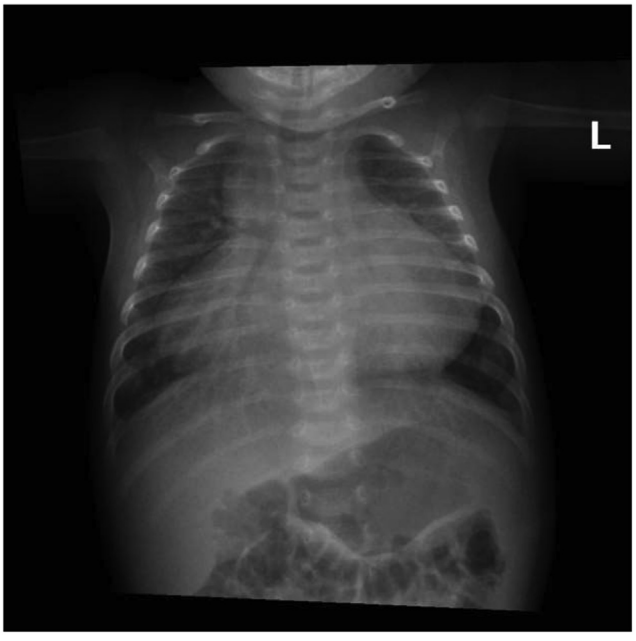

Figure 1 Chest X-ray, anteroposterior projection showing cardiomegaly with enlarged right atrial contour.

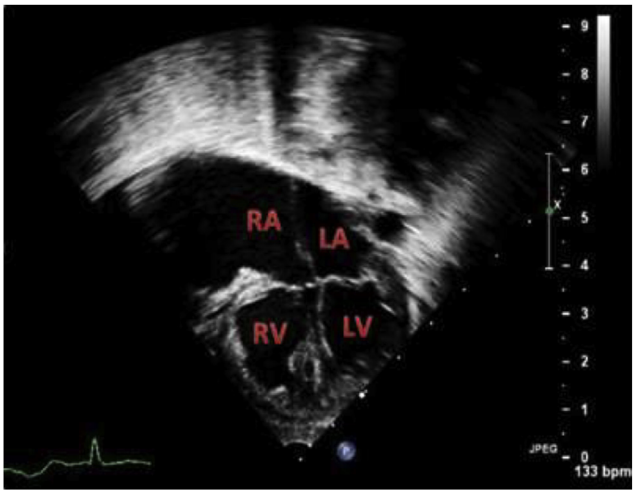

Figure 2 Transthoracic echocardiogram. The four-chamber view shows a massively dilated RA with normal position of the atrioventricular valves. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

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 left to right 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 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).

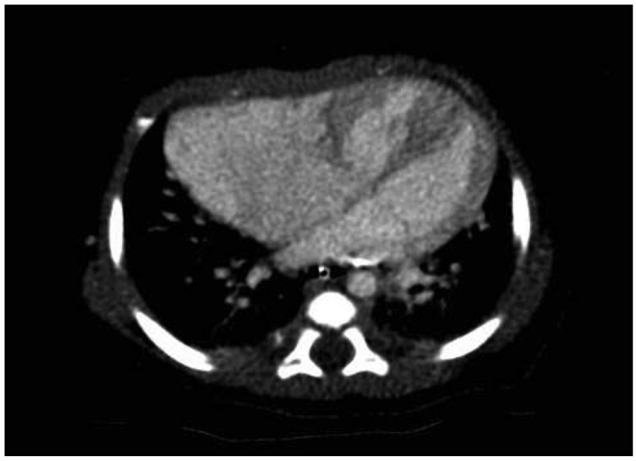

Figure 3 Enhanced axial CT scan of the chest showing a large dilatation of the right atrium and atrial appendage with a thin atrial wall.

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

Authors	Age and presentation	Treatment	Outcome
Mizui et al., 2001 [12]	Infant with ectopic atrial tachycardia	Surgical correction	Stable after surgery
Tejero-Hernández et al., 2012 [5]	Prenatal diagnosis	Medical treatment	Stable at 1-year, 6-months follow-up
Ishii et al., 2012 [13]	Prenatal diagnosis	Careful observation	Stable at 8-months follow-up
Lang et al., 2014 [9]	Prenatal diagnosis	Surgical correction	Stable at 2-months follow-up
Tunks et al., 2015 [2]	Prenatal diagnosis	Surgical correction	Stable at 4-months follow-up
Cardiel Valiente et al., 2016 (2 cases) [14]	Prenatal diagnosis Prenatal diagnosis	Medical treatment Careful observation	Gradual regression Stable throughout the follow-up

4. Conclusion

Right atrial appendage aneurysms can be diagnosed in asymptomatic newborns. 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

The authors have no conflict of interest to report.

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