Prenatal diagnosis of right atrial diverticulum and outcomes: a case report

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Case Report

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Abstract

Background

Right atrial diverticulum is a rare congenital condition which causes the right atrium enlargement. The squeals of this condition can vary from cardiac abnormalities to respiratory distress and systemic thromboembolism, hence Identifying these patients can prevent life-threatening outcomes. Prenatal diagnosis has the benefit of better following up and managing patients to prevent later subsequences.

Case presentation

Echocardiography of the neonate 3 days after birth showed a massive right atrium with a diverticulum measuring 2.09*2.27 cm connected laterally to the right atrium without any clot in it. A fibromuscular strand was seen at the entry of the diverticulum through the right atrium.

CT-Angiography 16 days after birth showed a massive right atrium with a diverticulum in the right hemithorax and confirmed the diagnosis.

The patient underwent on low dose aspirin therapy to prevent thromboembolism.

After 16 months the patient goes on well this condition without cardiac or respiratory symptoms and echocardiography showed the diverticulum size increased to 3.5*2.5 cm without any clot in it.Surgical resection has not proceeded yet because the patient has been asymptomatic until now.

Conclusion

Because of the rarity of this condition, management of these patients is highly dependent on the symptoms they show, and also early diagnosis can prevent further medical squeals. Low doses aspirin is suggested to prevent the formation of thrombosis. Surgical resection can be done in patients with serious cardiac or respiratory abnormality,

It is important to do not misdiagnosis this condition with a right atrial aneurysm which involves whole layers of the atrial wall. Although outcomes of both conditions are almost the same, using a proper term to establish an accurate diagnosis preferred.

Introduction

Right atrial diverticulum also known as by the other names such as the idiopathic dilatation of right atrium(IDRA)(1, 2), right atrial aneurysm(RAA)(3), or even aneurysm of the right atrial appendage(4, 5), is a rare a condition which most often diagnosed by chance in adults(2, 4), but the prenatal and antenatal diagnosis of this condition has been reported even more rarely.(1, 3)while IDRA or RAA defined as the isolated enlargement of the right atrium without the presence of any other cardiac lesions which can cause right atrium dilatation, a diverticulum is a fibromuscular strand entry of the right atrium and it is

important from this aspect that can be misdiagnosed with right atrial aneurysm which involves all layers(epicardium, myocardium, and endocardium) of the atrial wall, while it is better this two conditions to be differentiated from each other(1, 6)in the literature, this differential diagnostic border has not been clearly established and that's why we see these two different terms are used frequently instead of each other as we can see in Morrow et al report(7). Other misdiagnoses such as Ebstein's anomaly should be ruled out using fetal heart echocardiography(3, 8). Many cases presenting with right atrial enlargement are asymptomatic(4), however, long-term outcomes and prognosis should not be overlooked, hence it can lead to cardiac arrhythmia, tachycardia, atrial fibrillation, thromboembolism, and respiratory distress due to high pressure of the massive right atrium on the left bronchi and causing airway compression(3). Management of the right atrial enlargement is still controversial and varies widely from routine clinical follow up to anti-arrhythmic prophylaxis and surgical resection depended on the clinical presentation(3, 4).

Case Report

A 38-year old primigravid woman presented to our clinic at the 37th weeks of gestation for routine fetal heart echocardiography with the past medical history of using clomiphene and levothyroxine for infertility and hypothyroidism disorder respectively, which discontinued prior the pregnancy. fetal echocardiography showed a massive right atrium with a massive appendage. A male neonate delivered vaginally at the 39th weeks of gestation without any complication with the birth weight of 3.200 kg.

3-day old Neonate echocardiography showed a diverticulum within the entry of the right atrium wall measuring 2.09 × 2.27 cm located laterally (Fig. 1, overlaid in green).

As we mentioned definition of the diverticulum in the introduction, a fibromuscular strand is seen which connected to the superior surface of the right atrium and moves synchronously with heart movement and at the first sight it seems there is an extra cavity attached to the right atrium(vid.1)

There was a mild tricuspid regurgitation (PG = 18 mmHg), foramen oval was patent and ventricular septum and function reported normal, hence Ebstein's anomaly has been ruled out(vid.1)

A chest CT-angiography on the 16th day of the neonatal life revealed a massive right atrium in the right hemithorax space, a large wide-necked diverticulum of the right atrium is seen, no clot formation detected.

Interatrial and interventricular septum were intact and the main pulmonary artery and branches were normal(Fig. 3,4,5)

The patient is under observation and clinical follow-up on daily low dose aspirin for thromboprophylaxis.

Growth and other related factors such as oxygen saturation was normal and there wasn't any sign of cyanosis, thrombosis, arrhythmia, or respiratory distress.

Surgical resection has not been made yet, since the patient gets on well with this condition for 16 months after birth with a diverticulum measuring 3.5 × 2.5 cm.

Discussion

Right atrial enlargement is an idiopathic condition that has been reported from fetal life to the elderly(2, 3, 4). It is also known by other terms such as congenital aneurysm or diverticulum of the right atrium(2, 4, 7)Prenatal diagnosis has the benefit of identifying fetuses with cardiac abnormality(1, 3)and termination of the pregnancy have been proceeded in some cases(5). also, there were cases diagnosed with older ages while they were asymptomatic for a longer period (1, 3, 4, 6), the outcomes of some patients diagnosed with congenital right atrial aneurysm were accompanied by the life-threatening conditions, such as atrial tachycardia, thromboembolism, respiratory distress, and atrial arrhythmia(3)such conditions require more consideration and attention in patients follow up.

while we expected our presented case manifest with signs of arrhythmia, thromboembolism, or cardiac dysfunction in early ages of birth, even after 1 year and 6 months our patient didn't experience any of these conditions, it led us to an important point in differential diagnosis about this disease. with the focusing on the terminology, an atrial aneurysm is defined as a dilated atrium which involves whole layers of its wall, while in our case there was an entry across the atrial wall (which resembles the diverticulum's neck) with a fibromuscular strand stretched to the superior surface of the right atrial wall and this condition is more consistent with a diverticulum(Fig. 5)(1, 6)

Conclusion

Atrial diverticula have been reported from birth to adult life which many of them were asymptomatic. as we discussed it is important to differentiate a diverticulum from an aneurysm, but risks of both conditions should not be overlooked, hence both of them can cause fatal events.

The role of echocardiography in diagnosing and managing patients with right atrial enlargement either in fetal life or adult life is significant and undeniable. Using low doses of anti-coagulants agents such as aspirin is also useful for thromboprophylaxis.

Finally, surgical resection is suggested in cases with respiratory or cardiac symptoms.

Declarations

Ethics approval and consent to participate:

We assure that any identity information related to this patient remained confidential and we hided any name or code related to this patient in our images and figures. Our patient and his guardian consented to participate in this report.

Consent for publication:

Our patient and related guardian gave consent for publication and gave consent to provide supplementary data in this report.

Availability of data and materials:

Data sharing is not applicable to this article, as no datasets were generated or analyzed during the current study.

Competing interests:

The authors declare that they have no competing interests.

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Authors' contributions:

All authors contributed the same to provide this report and both authors confirm this report.

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Figures

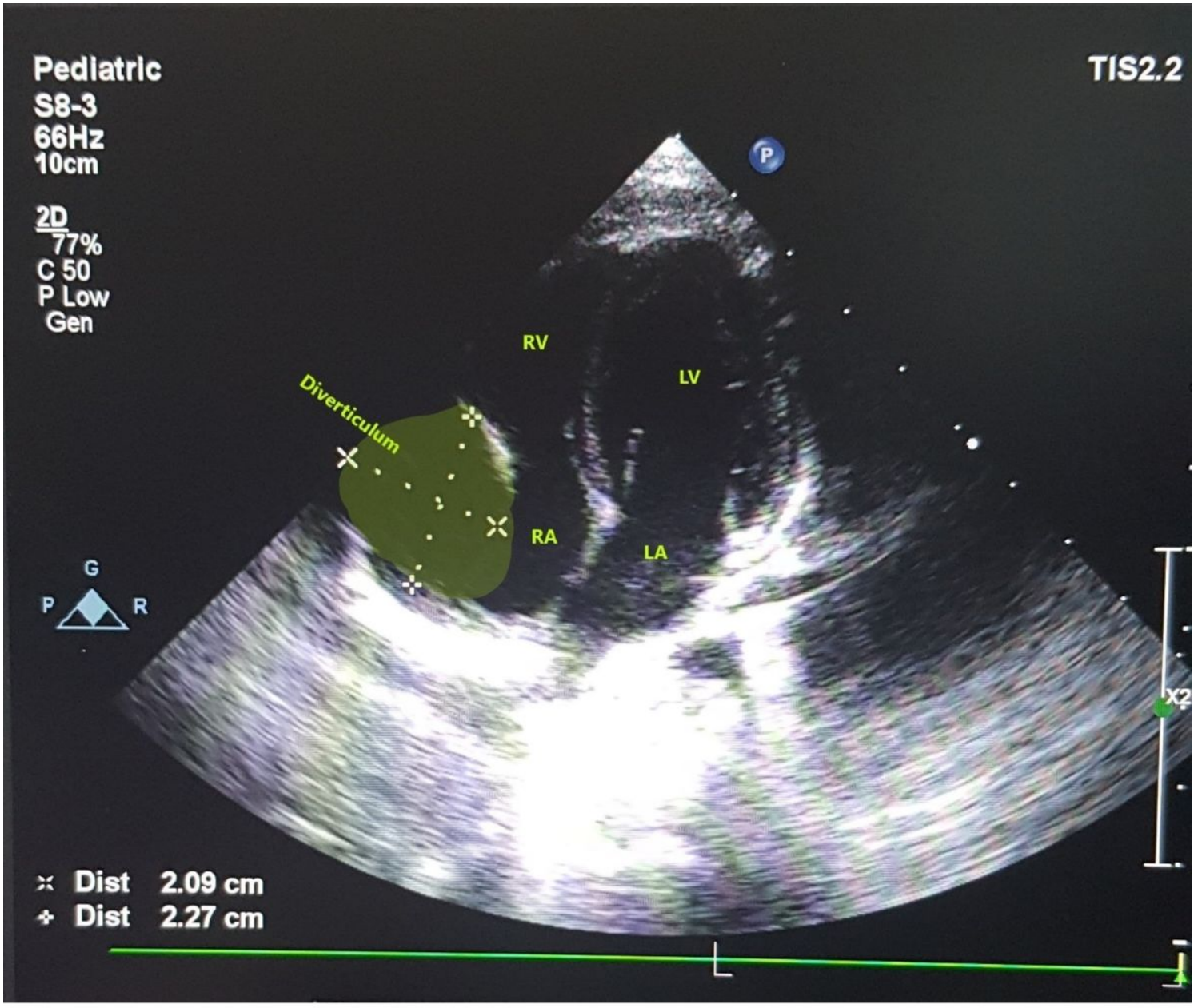

Figure.1-echocardiography of the 3 days old neonate's heart showed a diverticulum measuring 2.09*2.27 attached to the lateral wall of the right atrium.

Figure 1

echocardiography of the 3 days old neonate's heart showed a diverticulum measuring 2.09*2.27 attached to the lateral wall of the right atrium.

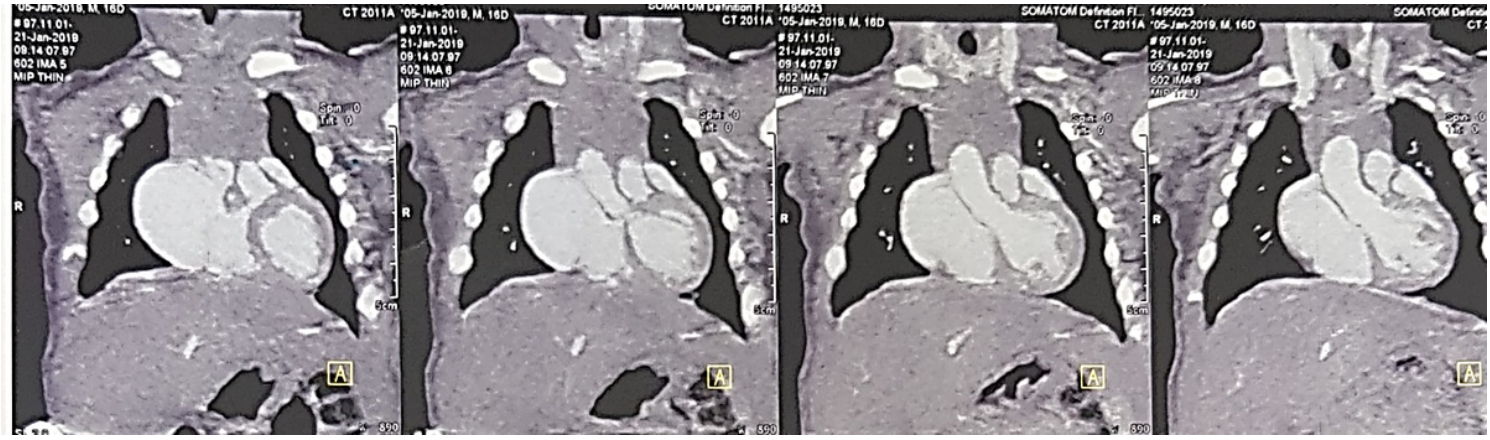

Figure 2

Coronal view CT-Scan of the neonate's heart.

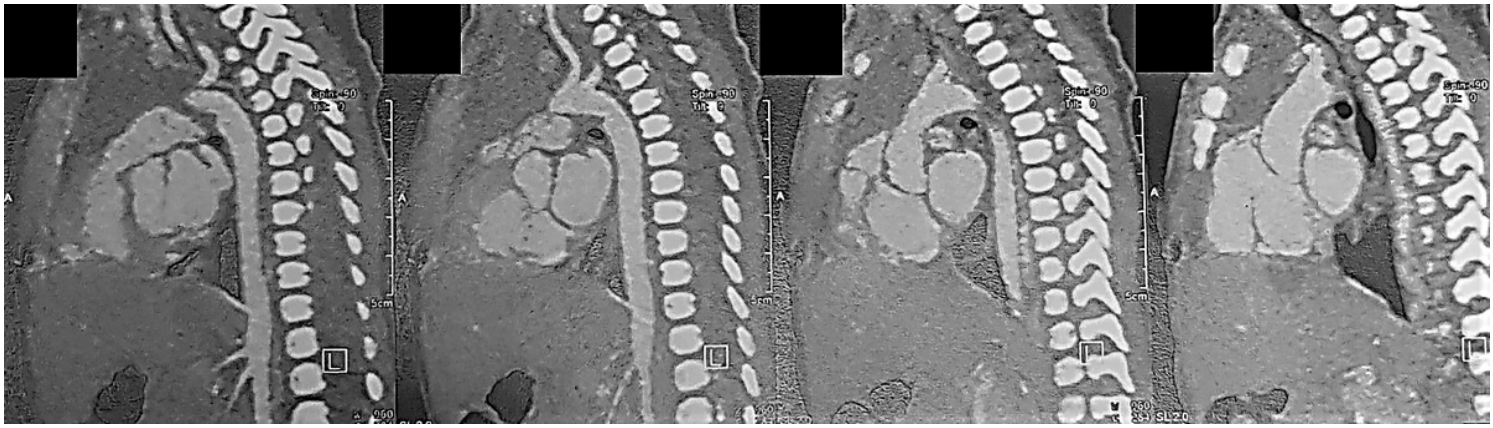

Figure 3

Sagittal view CT-Scan of the neonate's heart.

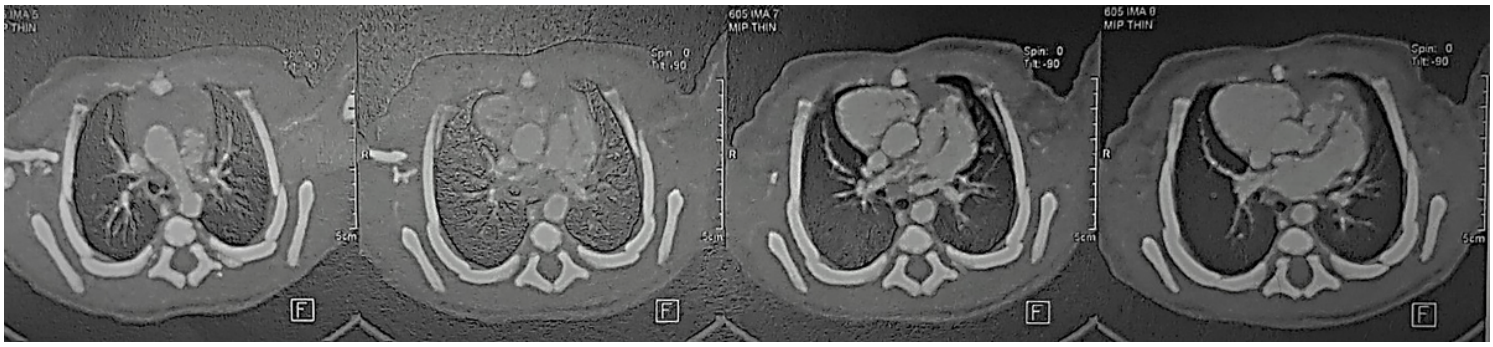

Figure 4

Axial view CT-Scan of the neonate's heart.

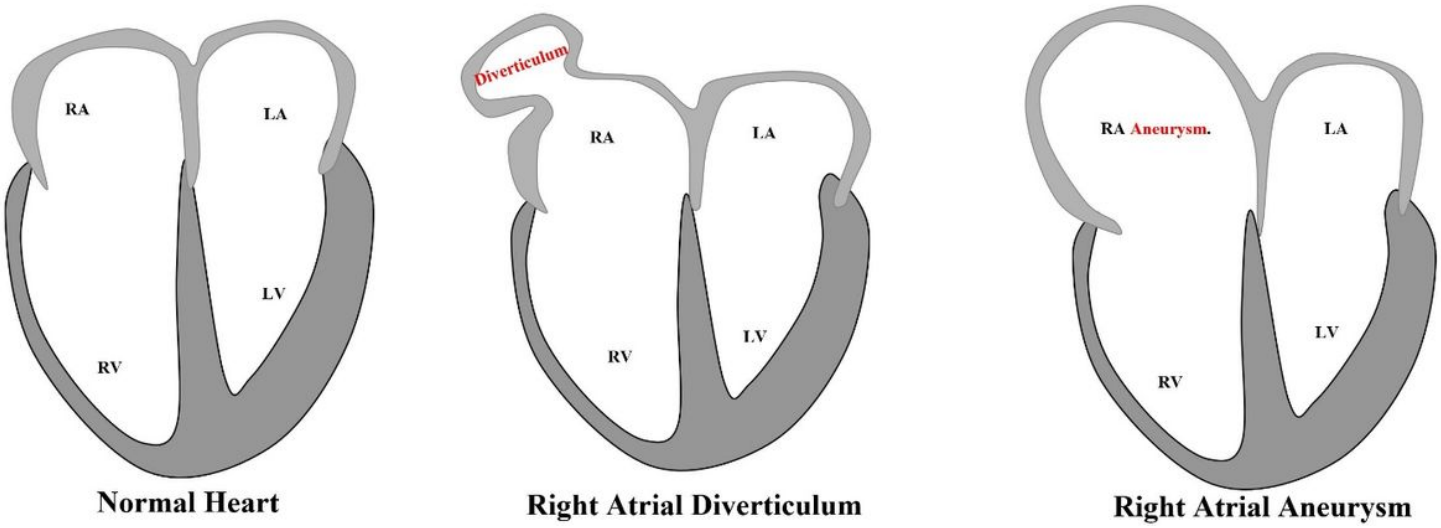

Figure 5

A schematic picture of a noraml heart, right atrial diverticulum. and right atrial aneurysm.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- vid1.mp4
- CAREChecklist.docx