**CARE Checklist**

1. **Title**: Prenatal diagnosis of right atrial diverticulum and outcomes: a case report
2. **Key Words**: right atrial enlargement-right atrial diverticulum-aneurysm-prenatal diagnosis
3. **Abstract(summary)**

**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.

1. **Introduction(detailed with references)**

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).

1. **Patient Information:**

We report a case diagnosed with right atrial diverticulum at the 37th weeks of gestation and then followed up after birth for 1 year and 6 months.

Prenatal fetal echocardiography revealed right atrial enlargement and neonate echocardiography at day of 3 confirmed prenatal diagnosis.

The patient was asymptomatic after birth and for preventing thrombosis low dose aspirin started and patient remained asymptomatic for 18 months.

Mother was a 38-years old primigravid woman with a history of using clomiphene and levothyroxine for infertility and hypothyroidism disorder respectively, which discontinued prior the pregnancy.

Patient did not have similar familial history of congenital cardiac abnormality in first- and second-degree relatives.

1. **Clinical Findings:**

Right atrial enlargement diagnosed at 37th weeks of gestation and echocardiography 3 days after birth and CT-Angiography 16 days after birth confirmed diagnosis of right atrial diverticulum.

Patient did not show any sign of cyanosis, thrombosis, arrhythmia, or respiratory distress while following up and remained asymptomatic for 18 months period.

1. **Timeline:**

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| **Time** | **Findings** |
| 37 weeks of gestation | Prenatal echocardiography diagnosed a large right atrium. |
| 39 weeks of gestation | Patient delivered vaginally |
| 3 days after birth | Patient echocardiography showed a right atrial diverticulum measuring 2.09\*2.27 cm. |
| 16 days after birth | CT-Angiography confirmed the diagnosis of right atrial diverticulum.  Low dose aspirin started for patient. |
| Serial echocardiography until age of 18 months | final echocardiography at the age of 18 months showed right atrial size increased to 3.5\*2.5 |

1. **Diagnostic Assessment:**

Fetal heart echocardiography performed at the 37th weeks of gestation and 3 days after birth echocardiography of neonate’s heart performed.16 days after birth a CT-Angiography was done.

The most important challenge in this case was misdiagnosing this condition with right atrial aneurysm which involves whole atrial wall layers and also there is no wide necked inlet attached to right atrium in an aneurysm, so establishing an accurate diagnosis and using a correct term should not be overlooked.

1. **Therapeutic Intervention**

To prevent thrombosis and clot formation patient started daily 100 mg aspirin consumption when diagnosis has been established and continued for 18 months since now.

Surgical resection has not been made since the patient was asymptomatic.

1. **Follow-up and Outcomes**

While following up patient was asymptomatic, serial echocardiography showed increasing the size of diverticulum from 2.09\*2.27 cm(day of 3) to 3.5\*2.5(month of 18th)

1. **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(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.

1. **Patient Perspective**

Our patient was asymptomatic for 18 months like any other healthy child, however role of aspirin in preventing thrombosis in such patients is very important, since clot formation and stasis can occur at any stage of time.

1. **Informed Consent**

Our patient and his guardian gave informed consent about any stage of follow up and treatment and also consented to share patient’s medical condition for writing this report.

All identity information about this patient protected and remained confidential.

**References**

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