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Idiopathic enlargement of the right atrium masking left atrial aneurysm in a neonate

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Brief Report

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Abstract

An idiopathic enlargement of the right atrium is an extremely rare cardiac malformation. There are no established guidelines for the management of this disease, especially concerning medical versus surgical therapeutic approach and the timing for an operation. We report in this case about a neonate that first was treated conservatively until the age of 5 month and finally got an operative resection of the aneurysm. After surgery, unexpected complications occurred. A second aneurysm in the left atrium was demasked. Furthermore, a progressive dilatation of both atrial chambers after resection required regular follow-up and ongoing evaluation of treatment.

An idiopathic enlargement of the right atrium, also called congenital right atrial aneurysm, is an extremely rare malformation of the right atrium. It is defined by an isolated dilatation of the right atrium in the absence of primary cause for right atrial dilatation, like atrial septal defects, Epstein's anomaly, anomalous pulmonary venous return, and lesions of tricuspid or pulmonary valves.[1] It has been first described and treated by surgical resection by Bailey[2] in 1955. Since then numerous cases have been published in literature, but there are only a few systematically reviews, such as the work of Binder et al. (2000)[3] and recently by Jing Zhang et al. (2021).[4] Nevertheless, it remains outside of clinical routine, and there is no standardised management of these disease. We report in this case about a neonate that first was treated conservatively until the age of 5 month and finally got an operative resection of the aneurysm. After the intervention, a left atrial aneurysm was demasked and made a second operation necessary. To the best of our knowledge, a congenital right atrial aneurysm masking a left atrial aneurysm has never been described in literature previously.

Case

We describe a male neonate with congenital right atrial aneurysm that has been detected prenatally during routine fetal ultrasound screening. The postnatal period was uneventful. The baby showed normal heart rate, blood pressure, and oxygen saturation. To prevent right atrial thrombosis and embolic complication, an anticoagulant therapy with heparin was directly started after birth.

The electrocardiogram showed an ectopic low atrial rhythm and P-pulmonale with a normal frequency. On a Holter-electrocardiogram, no signs of supraventricular tachycardia or other pathological arrhythmia were observed. Postnatal thoracic X-ray revealed cardiomegaly with a cardio-thoracic ratio of 0.82 (Fig. 1c). Transthoracic echocardiography showed a massif dilatation of the right atrium of 10 cm^2 and a small atrial septal defect II with an insignificant left-right-shunt (Fig. 2a). Besides a small spur in the descending aorta with no haemodynamic relevance and a bicuspid aortic valve, there were no other structural anomalies. The other heart chambers, especially the left atrium, were normal-sized, and the atrioventricular valves were competent without displacement or regurgitation. Cardiac MRI showed the dilated right atrium with 14.11 cm^2 (Fig. 1a). Due to the huge right atrial aneurysm, dorsal pulmonary structures and the pulmonary veins appeared compressed. Especially, both lower pulmonary veins appeared small with a mild functional stenosis of the left lower pulmonary vein.

To reduce potential perioperative complications of extracorporal circulation in neonates and due to the fact that the child was asymptomatic and had no signs of heart failure or severe arrhythmia, we decided to pursue a conservative approach until the age of 4-5 month. The child received aspirin for thrombo-embolic prevention and bisoprolol as anti-remodelling-therapy. The operative resection was performed at the age of 5 month. At this time, the child showed a good development and, apart from a moderate tachycardia, no clinical symptoms, but the right atrial aneurysm has progressed to 23 cm^2. Intraoperative, the right atrium impressed as giant aneurysm that filled the hole mediastinal space. (Fig. 1b). Visually, it could easily be distinguished between aneurysm and atrial myocardial tissue. The aneurysm was resected and the ASD II closed. The histopathological analyses of the aneurysmatic tissue revealed a thin predominantly fibrotic tissue with scant of myocytes.

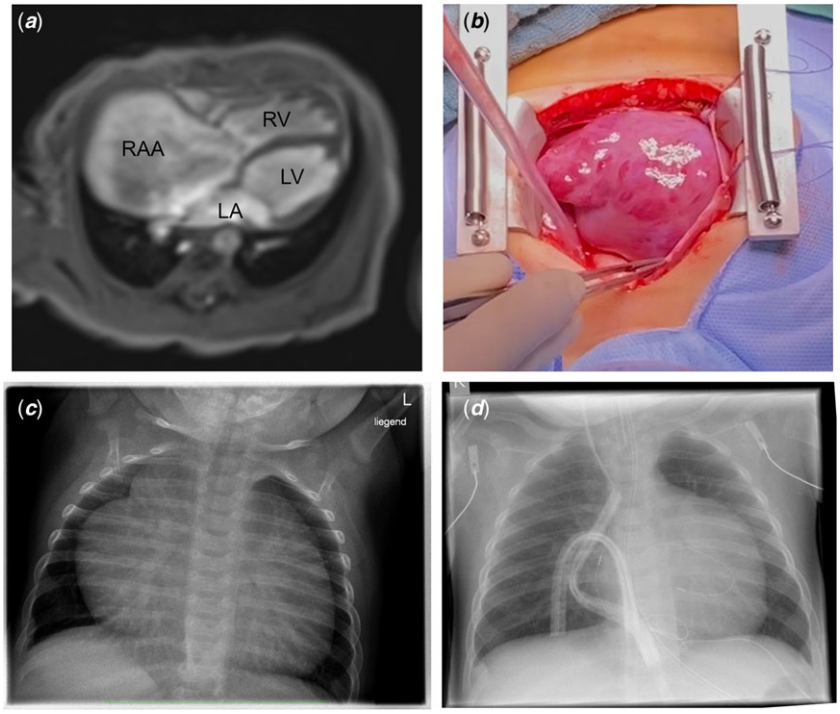

Figure 1. (***a***) Cardiac MRI demonstrating the large right atrium, (***b)*** intraoperative the RAA is filling the hole situs, (***c***) preoperative X-ray shows a CTR of 0.82, (***d***) and a significant reduction of the cardiac size after resection. LA = left atrium; LV = left ventricle; RAA = right atrial aneurism; RA = right atrium; RV = right ventricle.

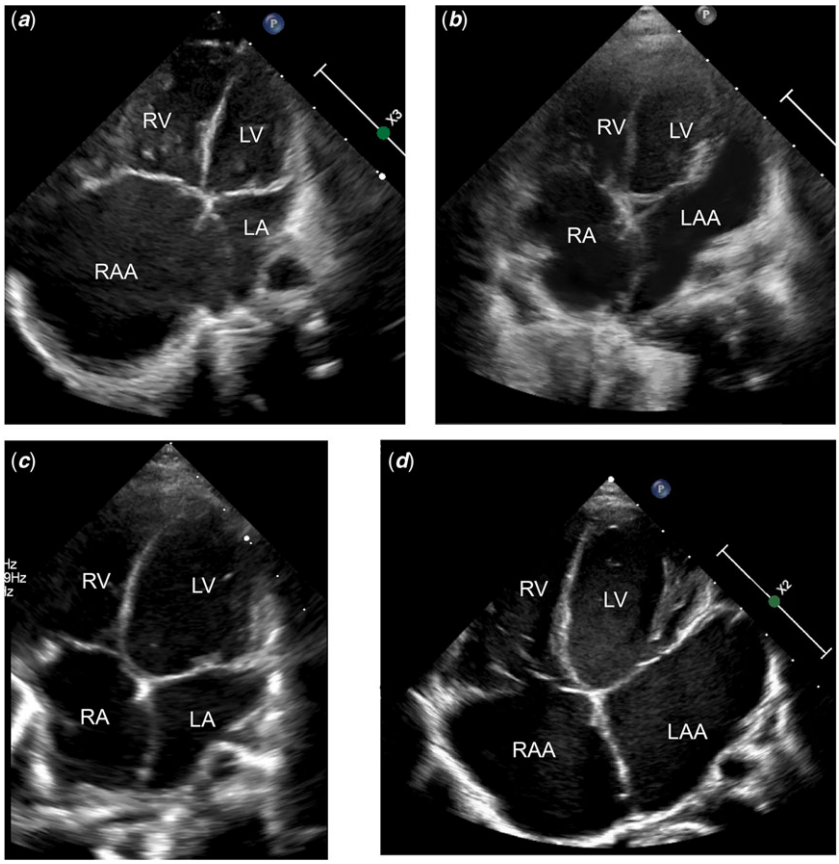

Figure 2. (***a***) An apical 4-chamber view in transthoracic echocardiography shows a giant right atrium. (***b)*** After surgical resection of the RAA, a left atrial aneurysm was demasked. (***c***) After second surgery, the echocardiography shows normal-sized right and left atrium. (***d***) Progress of both atrial chambers in a 10 month follow-up. LAA = left atrial aneurism; LA = left atrium; LV = left ventricle; RAA = right atrial aneurism; RA = right atrium; RV = right ventricle.

Echocardiography a few hours after surgery showed a normal-sized right atrium and unexpectedly a 9.0 cm^2 large aneurysm of the left atrium (Fig. 2b). To specify this finding and planning, further therapy an MRI was performed, where an aneurysm of the left auricle was confirmed. Seven days after the first surgery, a second operation was performed on beating heart to resect the left atrial aneurysm. The control thoracic X-ray showed a normal-sized heart with a cardio-thoracic ratio of 0.5. (Fig. 1d). Post-operative

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transthoracic echocardiography revealed good biventricular function and slight dilatation of right (6.3 cm^2) and left atrium (5.5 cm^2), as well as mild mitral and tricuspid regurgitation (Fig. 2c). Electrocardiogram showed an alternating sinus and junctional rhythm with stable hemodynamic.

At follow-up examination, 3 weeks later the child was in good clinical condition, without any symptoms of heart failure or arrhythmia, but the echocardiography again showed a progress of right (8.4 cm^2) and left aneurysm (8.9 cm^2) dilatation. The latest follow-up 10 month after the second operation demonstrated a further progression of the left atrial size to 14,1 cm^2 (Fig. 2d), a moderate mitral regurgitation and recurrent focal atrial tachycardia (max. 220 bpm). Due to the ongoing progression of left and right atrial aneurysm, a genetic diagnostic was initiated. No genetic disorder affecting connective tissues or causing cardiomyopathy could be found. An advanced ultrastructural examination showed a disorder of myofibrillar architecture concerning the z-line and intercalated discs in cardiomyocytes.

Discussion

Idiopathic enlargement of the right atrium is an extremely rare heart defect of unknown aetiology. It can be diagnosed at any time in life, often by incidental finding on chest X-ray or electrocardiogram abnormalities.[5] A growing number of diagnosis at fetal or neonate age argue for a congenital cause of the aneurysm. So far the majority of right atrial aneurysms seem to be isolated cases, and a hereditary component was described by Jenni et al.[6] and Kurz et al.[7] Most common complications are supraventricular arrhythmias such as atrial fibrillation or supraventricular tachycardia and thrombo-embolism.[1,3–5] Only in the minority of patients a continuous progress of right atrial dilatation is described.[5] Although a number of cases have been reported since 1955, there are only a few systematic reviews and long-term follow-ups concerning this pathology. This still makes the management of congenital right atrial aneurysm an individual decision, especially concerning medical versus surgical therapeutic approach and the timing for an operation. For decision-making, the work of Binder et al.[3] and Zhang et al.[4] may be helpful. Binder analysed 60 and Zhang 153 cases (70 of them children) of right atrial aneurysm. Binder showed that patients that present any symptoms (mostly arrhythmia) will benefit of a surgical treatment, while asymptomatic patients – 48% of the analysed cases – should be treated conservatively. Zhang analysed that less than half of children with diagnosed RA-aneurysm showed complications at diagnosis, while most adult patient presented with complications. According to Zhang, a reason for this could be attributed to the short course of pathology in children. In line with Binder, Zhang identified any symptoms, mainly arrhythmia as a risk factor for adverse events and poor outcome.[4] Consequently, a surgical intervention might promise success for symptomatic patients. However, the best approach for asymptomatic patients is still controversial. In asymptomatic children, a prophylactic resection of right atrial aneurysm may be considered, especially because of the potential risk of thrombo-embolic complications.[8] Further research, based on long-term follow-ups, and longitudinal studies are necessary to develop guidelines. Currently, management of isolated right atrial aneurysm remains an individual decision based on the individual risk assessment.

In our case, the resection of right atrial aneurysm demasked a left atrial aneurysm that cannot be detected in echocardiography or cardiac MRI before. An explication for this could be that huge size of the right atrial aneurysm compressed the left atrial aneurysm. After resection, there was enough space to allow for unfolding in the thorax and a second operation deemed necessary. Furthermore, we observed an ongoing progression of right and left atrial dilatation in the early post-op follow-ups, indicating that regular follow-up is mandatory. Also, an advanced cause research including a genetic diagnostic and an advanced histopatholoigical examination of the resected tissue is required. Even if the genetic analysis in this patient did not show a mutation that could explain the nature of biatrial aneurysm, a major underlying disease must be considered. Due to this case, we raise the question of whether defects in genes that affects the early embryonal heart could be causing this disease. Specialy progenitor cells from the second heart field that generates distinct cardiac chambers could be affected.[9] Whether underlying genetic defects for atrial dilatation can be determined is reserved for future research.

Conclusion

Although most authors suggest surgical resection of right atrial aneurysm as preferred treatment option, unexpected complications can occur. In our case, the demasked left atrial aneurysm and a progressive dilatation of both atrial chambers after resection required regular follow-up and ongoing evaluation of treatment. The nature of congenital atrial aneurysm currently remains unknown. Major underlying genetic defects for atrial aneurysm must be considered for optimal choice of the therapeutical approach. In the absence of established guidelines, the management of patients with congenital atrial dilatation still remains an individual decision.

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