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Brief Report

Aorta–right atrial tunnel closure using the transcatheter technique: a case of a 3-year-old child

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Abstract Aorta–right atrial tunnel is a rare congenital defect. If it is not treated, critical complications can occur. Surgical closure is the treatment of choice, but with increasing experience in transcatheter techniques and the introduction of new occluder devices the percutaneous technique is now a successful modality for closure. In this paper, we report the case of a 3-year-old child with aorta–right atrial tunnel whose defect was closed with the vascular plug 4 device.

Keywords: Tunnel; vascular plug; transcatheter closure; amplatzer

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Aorta–right atrial tunnel is a rare congenital connection.[1–3] Until now, only 17 cases have been reported in the literature and most of these were adults. In this paper, we describe our experience of a newly diagnosed rare defect, aorta–right atrial tunnel, and its successful closure with an Amplatzer® vascular plug 4 device (St. Jude Medical Inc., St. Paul, Minnesota, USA).

Case

An 8-month-old girl whose arterial duct was closed in another centre was referred to our clinic for continuing murmur on the right upper sternal border. There was no pathological electrocardiography finding, and the cardiothoracic ratio was in the normal range. A continuous flow pattern was present from the left coronary sinus through a tunnel to the right atrium and an apical small ventricular septal defect was detected on echocardiography. A 30 millimetres of mercury pressure gradient was measured at the orifice of the tunnel.

An angiocardiography was performed and an apical small ventricular septal defect was detected; the pulmonary artery peak pressure was measured as 33 millimetres of mercury. The Qp/Qs ratio was 1.88. Aortic root angiography revealed the normal right and left main coronary arteries and branches, but also a large tortuous tunnel originating from the left coronary sinus measuring approximately 4 centimetres in length and 5.5 millimetres in diameter passing posterior to the aortic root and terminating near the right atrial–superior caval vein junction (Fig 1). However, there was no coronary branch originating from the tunnel. After placing a long multipurpose catheter to the tunnel an Amplatzer® vascular plug 4 device advanced inside this catheter. This device was selected because of its flexibility and small profile, allowing it to reach more distal vessels than other plugs. The tunnel was closed with this device (Fig 2). Control aortic root angiography revealed that the vascular plug was in a stable position at the entrance of the tunnel and a minimal shunt was observed inside the tunnel (Fig 3). No continuous murmur was heard on the right upper sternal border on the first day, 15th day, and first month check-ups. In addition, residual shunt was not detected in the tunnel on transthoracic echocardiography. Periodic follow-up of the patient continues in our unit, and the control angiography performed after 6 months of the procedure revealed no residual shunt (Supplementary Movie).

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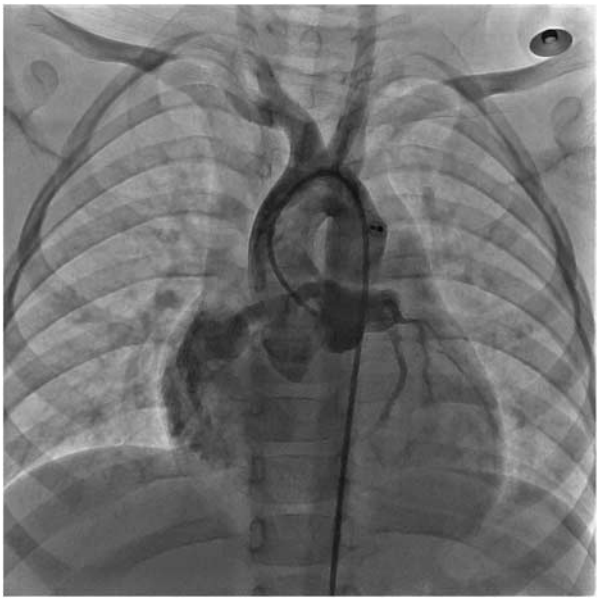

Figure 1.
An abnormal tunnel originating from the aneurysm of the left coronary sinus to the right atrium and the normal anatomy of the coronary artery.

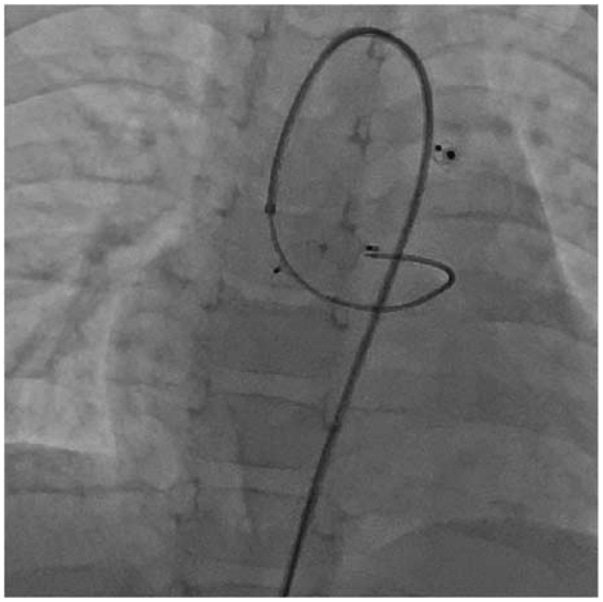

Figure 2.
An image of the 8 millimetre Amplatzer® vascular plug 4 in the tunnel.

Discussion

Congenital aorta–atrial tunnel, first described by Coto et al[1] in 1980, is an extracardiac congenital communication between the right atrium and the aorta. The most postulated mechanism for aorta–right atrial tunnel is aortic elastic lamina media deficiency.[4] The term aorta–atrial fistula is also used especially for secondary cases. It is hard to discriminate tunnel from coronary–cameral fistula. Generally, a tunnel differs from fistula in that it does not have myocardial branches.[2] In addition, the differentiation between a tunnel and sinus Valsalva aneurysm must be made. A tunnel originates above the supra-aortic ridge, whereas sinus Valsalva aneurysm is located under the ridge.[5,6] In our case, normal coronary artery anatomy was revealed by angiocardiography and there were no side branches originating from the tunnel. Therefore, the presence of normal major coronary arteries and absence of any small myocardial coronary branches from the tunnel argues against the structure being a coronary–cameral fistula and supports the diagnosis of aorta–right atrial tunnel.

Volume overload may happen in both ventricles if there is a critical shunt from left to right, and as a consequence exercise dyspnoea, tachycardia, chest pain, and recurrent pulmonary infections may be seen. Moreover, patients may be asymptomatic.[4] Our patient did not have any obvious symptoms and most of the cases in the literature were asymptomatic. Except for one of the published cases, all of the patients had a continuous murmur on the right side of the sternum, as was the case in our patient.

Although aorta–right atrial tunnel can be determined by transthoracic echocardiography, diagnosis may be confirmed by aortography and selective coronary angiocardiography. In our case, colour Doppler demonstrated a tunnel extending from the left aortic sinus to the right atrium.

Left coronary sinus aneurysm is the rarest type of coronary sinus aneurysms (1%), and when it occurs it can lead to rupture in adjacent structures such as the left ventricle, pulmonary artery, myocardium, and epicardium.[7] In our case, there was a communication originating from the left coronary sinus and passing behind the aorta to the right atrium. Hence, this communication was not thought to be rupture of a sinus Valsalva aneurysm.

Even though patients are asymptomatic, when the diagnosis is made it should be closed as soon as possible. The reason for this is to avoid the potential risk of emboli, spontaneous rupture thrombosis, aneurysm formation, infective endocarditis, pulmonary vascular disease, coronary failure, aortic insufficiency, and calcification on the wall of the tunnel.[1,8] In addition, the surgical mortality risk increases for the patients who are not treated until advanced ages.

Treatment options include transcatheter closure, ligation of the tunnel under controlled hypotension, and surgical closure under total cardiopulmonary bypass.[5] The transcatheter closure technique is safer because of the lower risk of artery, vein, and nerve injuries, infection problems, rhythm problems,

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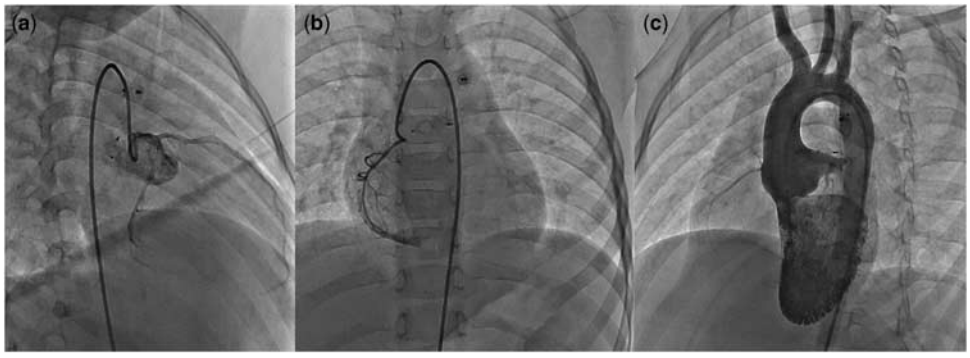

Figure 3.
*(**a**): Left coronary, (**b**): Right coronary, (**c**): Left ventricular angiographies after tunnel closure.*

hypoxia, and cosmetic problems. To the best of our knowledge, 17 cases have been reported in the literature; two of them were closed by transcatheter technique – both were paediatric patients – 14 of them were closed by surgery, and one of them was accepted for clinic monitoring. Sreedharan et al[5] were the first to report a case in which the tunnel was embolised using two units of coil with transcatheter technique in an 11-year-old patient in 2006. Mahesh et al[9] reported a case of transcatheter closure of an aorta–right atrial tunnel using coil for a 4-day-old baby. We presented a case of this rare disease and percutaneous application – successful closure of the tunnel using an Amplatzer® vascular plug 4 with the transcatheter technique. Using coil may be preferred because it is an inexpensive and simple technique, but the length and diameter of the tunnel led us to select the Amplatzer® vascular plug 4, because the coil may have migrated to the right atrium because of the large distal end of the tunnel. In addition, the design and the softness of the Amplatzer® vascular plug 4 was suitable for this particular case.

Conclusion

Aorta–right atrial tunnel is a rare congenital lesion with an unknown aetiology. Owing to the possible complications, it should be closed after certain diagnosis. In addition to citing two other cases in the literature, which have been closed using the transcatheter procedure, we present the first case, which was completely closed with the Amplatzer® vascular plug 4 device. Thus, we aim to highlight this rare disease with our experiences on its diagnosis and management.

Supplementary materials

For supplementary material referred to in this article, please visit http://dx.doi.org/doi:10.1017/S1047951112001151

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