Two cases of aorto-right atrial tunnel: clinical presentation, imaging and percutaneous closure

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Abstract Communication between the aortic sinus and a cardiac chamber is a rare anomaly that can be diagnosed in children and young adults. We describe two cases with a tunnel between the aortic sinus and right atrium, discuss diagnostic modalities, and review the literature on this anomaly.

Keywords Aorto-right atrial tunnel · Congenital heart disease

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Introduction

With the improvement of non-invasive imaging and percutaneous techniques, complex and rare congenital cardiovascular conditions can nowadays be treated in the catheterisation laboratory by a multidisciplinary team. Communication between the aortic sinus and a cardiac chamber is a rare anomaly, which can be diagnosed in children and young adults. Aorto-right atrial tunnel or aorto-right atrial fistula (ARAT or ARAF) was first described in 1980 by Otero-Coto and colleagues,[1] and only a few cases have been reported in the literature. It has been defined as ‘an abnormal tubular extracardiac communication between the ascending aorta and the right atrium’.[2–6] This condition is distinguished from a coronary-cameral fistula because the origin is separated from the ostium of the coronary artery and because they may also arise from the non-coronary sinus.[7, 8] These communications may present as an isolated anomaly, or coincide with other congenital cardiac conditions. Clinical presentation may vary from completely asymptomatic, when the diagnosis is established by the presence of a loud, continuous cardiac murmur, or symptoms related to long-standing volume overload of both ventricles, such as shortness of breath or chest pain. We describe two cases recently diagnosed in our centre, along with diagnostic modalities and treatment options.

Case 1

A 23-year-old woman from Guinea, West Africa, was referred to our institution with a history of heart murmur since infancy. Over the past few years, she had become progressively

Fig. 1 Echocardiographic transthoracic 4-chamber view with colour Doppler, showing continuous flow inside the tunnel with turbulent flow at the connection with the right atrium

symptomatic with shortness of breath on exertion and palpitations. Several months before presentation she started experiencing continuous chest pain that increased during exercise. She had also experienced several episodes of syncope on exertion. Physical examination revealed a normal blood pressure but a resting heart rate of 105 beats/min. There was a loud, continuous grade 3 murmur midsternally. Chest X-ray showed marked cardiomegaly (cor-thorax ratio 0.57) and hilar bulging. In the four-chamber view, transthoracic echocardiography showed a large tubular structure behind the aorta running from left to right and draining into the right atrium (Fig. 1). There was continuous flow in the tunnel as evidence of a large left to right shunt and turbulent flow on the colour Doppler images, where the tunnel connected to the right atrium (Fig. 1). Dilatation of both the left and right ventricle was seen with preserved systolic function. Cardiac computed tomography (CT) scan provided suboptimal image quality due to the tachycardia, but a large tunnel was evident originating from the left coronary sinus with an approximate diameter varying from 10 mm to 17 mm, running posterior of the aorta and connecting to the right atrium. Cardiac catheterisation and angiography showed a large aorto-right atrial tunnel originating from the left coronary sinus (Fig. 2). The ostium of the left coronary artery was normal and visible high in the sinus, clearly separated from the entrance of the tunnel. The right coronary artery was normal. Aortic pressure and the pressure in the proximal tunnel was 115/75 mmHg, right ventricular pressure was 40/5 mmHg and mean right atrial pressure was 6 mmHg, demonstrating a clear pressure separation between the tunnel and the right atrium. The left to right shunt was quantified at 2:1.

A second procedure was performed under general anaesthesia with vascular access through the right femoral artery and vein. The tunnel was selectively engaged from the aorto using a 6F AL2 Amplatzer guiding catheter and a guideliner

Fig. 2 Angiographic still frames, showing the relation of the entrance of the aorto-right-atrium-tunnel (ARAT) with the left coronary sinus, the left coronary artery and the continuation of the tunnel towards the right atrium. **a** frontal view. **b** lateral view

extension. A 300 cm 35" Terumo wire was snared in the right atrium and exteriorised through the venous sheath (Fig. 3). An 8F Amplatzer guiding catheter was subsequently advanced from the right atrium into the tunnel and a 16 mm Amplatzer vascular plug (AGA Medical Corporation, Plymouth, USA) was deployed, closing the tunnel (Fig. 4a). However, the next day the vascular plug appeared dislodged from its position towards the right atrium, reopening the tunnel. During a third procedure using the same technique, an Amplatzer ASD closure device 16 mm (AGA Medical Corporation, Plymouth, USA) was positioned proximal to the vascular plug, closing the tunnel (Fig. 4b). Eighteen months after the procedure, the patient is still asymptomatic, her resting heart rate is 90 beats/min, and no murmur can be heard.

Case 2

A 3-year-old boy was first seen at the paediatric department at the age of 1 year because of mild signs of growth retardation below 2 standard deviations of the normalised growth curve. The habitus was normal, there were no signs of infection or malnutrition and his food intake and motoric development were normal. At the age of two, a continuous cardiac murmur was heard at the parasternal border in the third right intercostal space. The patient was asymptomatic, and no shortness of breath on exertion was noted. Echocardiography showed situs solitus, atrioventricular and ventricular-arterial concordance, with a normal left and right ventricular function. A large left to right fistula was seen, which was initially diagnosed as a left coronary artery-right atrium fistula. There was no coarctation and the ductus botalli was closed. Cardiac catheterisation and

Fig. 3 A guiding catheter was advanced via the aorta to the entrance of the tunnel, a wire was passed through the tunnel and snared in the right atrium. Subsequently, a large guiding catheter was passed from the right atrium into the tunnel and the device was positioned

Fig. 4 **a.** Shortly after the first device placement, the flow through the tunnel was absent. **b.** After placement of the second device, the next day, again the tunnel was closed

angiography at the age of 3 showed a large aorto-right atrial tunnel originating from the left aortic sinus, running posteriorly

Fig. 5 Angiographic image of case 2, demonstrating a similar aorto-right-atrial tunnel connecting the left coronary sinus to the right atrium with a posterior tunnel

and connecting to the right atrium (Fig. 5). There were normal left-sided and right-sided pressures, Qp:Qs was measured at 1.2. The coronary arteries were normal. As the patient was asymptomatic, he will be followed up at the outpatient clinic. Percutaneous closure of the tunnel will be scheduled at a later time point, depending on his symptoms and growth retardation, but preferably this will not be performed in the next few years.

Discussion

Our two cases of aorto-right atrial tunnel represent the wide spectrum of this congenital condition, ranging from an asymptomatic 3-year-old boy with a small left-to-right shunt to a young adult with severe symptoms and a large shunt. In both patients, the condition was characterised by a loud continuous murmur at the parasternal border. It is a rare condition, the first case was reported in 1980 and a review of the literature revealed a total of just 20 cases. Suitable imaging modalities include transthoracic and transoesophageal echocardiography, CT scanning, and cardiac catheterisation. These aorto-right atrial tunnels are separated into two types, running either posteriorly (as in both our cases) or anteriorly. The embryological background and cause of the anomaly is unclear. A possible explanation of this condition may be a congenital deficiency of the elastic lamina in the aortic media or confluence of coronary sinus. Most reported cases underwent surgical closure of the tunnel, either by direct closure of the right atrial entrance during cardiopulmonary bypass, or by simple ligation of the tunnel without cardiopulmonary bypass. Recently, several patients were reported to have undergone percutaneous closure of this anomalous tunnel with coils, vascular plugs and duct occluders. Closure of the tunnel is recommended once the diagnosis is established as it may lead to cardiac failure, aneurysm formation, spontaneous rupture and infective endocarditis. To our knowledge, this is the first

report of successful closure of an aorto-right atrial tunnel with a combined vascular plug and ASD closure device. The long-term follow-up of percutaneous closure of ARAT anomalies is largely unknown [9–11]. Our patient was free of symptoms after 18 months. In addition, the value of tunnel closure in asymptomatic young children with a small left to right shunt is also unclear, but ‘watchful waiting’ remains indicated.

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