Case Reports

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Aorta–Right Atrial Tunnel

Aorta–right atrial tunnel is a vascular channel that originates from one of the sinuses of Valsalva and terminates in either the superior vena cava or the right atrium. The tunnel is classified as anterior or posterior, depending upon its course in relation to the ascending aorta. An origin above the sinotubular ridge differentiates the tunnel from an aneurysm of the sinus of Valsalva, and the absence of myocardial branches differentiates it from a coronary–cameral fistula. Clinical presentation ranges from an asymptomatic precordial murmur to congestive heart failure. The embryologic background and pathogenesis of this lesion are attributable either to an aneurysmal dilation of the sinus nodal artery or to a congenital weakness of the aortic media. In either circumstance, progressive enlargement of the tunnel and ultimate rupture into the low-pressure right atrium could occur under the influence of the systemic pressure.

The lesion is diagnosed by use of 2-dimensional echocardiography and cardiac catheterization. Computed tomographic angiography is an additional noninvasive diagnostic tool. The possibility of complications necessitates early therapy, even in asymptomatic patients or those with a hemodynamically insignificant shunt. Available treatments are catheter-based intervention, external ligation under controlled hypotension, or surgical closure with the patient under cardiopulmonary bypass.

Herein, we discuss the cases of 2 patients who had this unusual anomaly. We highlight the outcome on follow-up imaging (patient 1) and the identification and safe reimplantation of the coronary artery (patient 2). ***(Tex Heart Inst J 2010;37(4):480-2)***

Aorta–right atrial (RA) tunnel is a vascular channel with its origin in one of the sinuses of Valsalva, a tortuous course anterior or posterior to the ascending aorta, and termination in the superior vena cava or in the RA. Herein, we discuss the diagnosis, treatment, and follow-up evaluation of 2 patients with this lesion who were treated at our institute: an 11-year-old girl who had a posterior tunnel and a 24-year-old man who had an anterior tunnel.

Case Reports

Patient 1. In December 2006, an 11-year-old girl presented with a 2-year history of worsening exertional breathlessness and palpitations. Cardiovascular examination revealed a hyperkinetic precordium and a continuous murmur at the right 2nd and 3rd intercostal spaces. Electrocardiography showed sinus rhythm, a normal axis, and right ventricular hypertrophy with a strain pattern. Chest radiography revealed a cardiothoracic ratio of 0.60.

Two-dimensional (2D) echocardiography showed a retroaortic tunnel from the left aortic sinus to the junction of the superior vena cava (SVC) and the RA. Cardiac catheterization revealed a Qp/Qs ratio of 1.8:1. Coronary angiography showed normal coronary arteries and a retroaortic tunnel from the left aortic sinus to the SVC–RA junction (Fig. 1A). There were no myocardial branches from the tunnel. Computed tomographic (CT) angiograms delineated the extracardiac anatomy (Fig. 1A inset and Fig. 1B). In view of the patient's symptoms and the substantial left-to-right shunt, surgical treatment was recommended.

Intraoperatively, the tunnel was seen to course along the antero-inferior aspect of the right pulmonary artery and to terminate on the posteromedial aspect of the dilated SVC–RA junction. An attempt to loop the tunnel under controlled hypotension failed, and elective cardiopulmonary bypass (CPB) was instituted. Initial cardiac arrest was achieved by means of retrograde coronary sinus cardioplegia. The SVC–RA junction was opened and the internal openings were identified. Noted upon exploration of the aortic root were moderate dilation of the left aortic sinus, a competent trileaflet aortic valve, and close juxtaposition of the aortic ostia of the left main coronary artery and the tunnel. To prevent injury to the ostium of the left coronary artery, the aortic orifice of the tunnel was left undisturbed. The RA openings were

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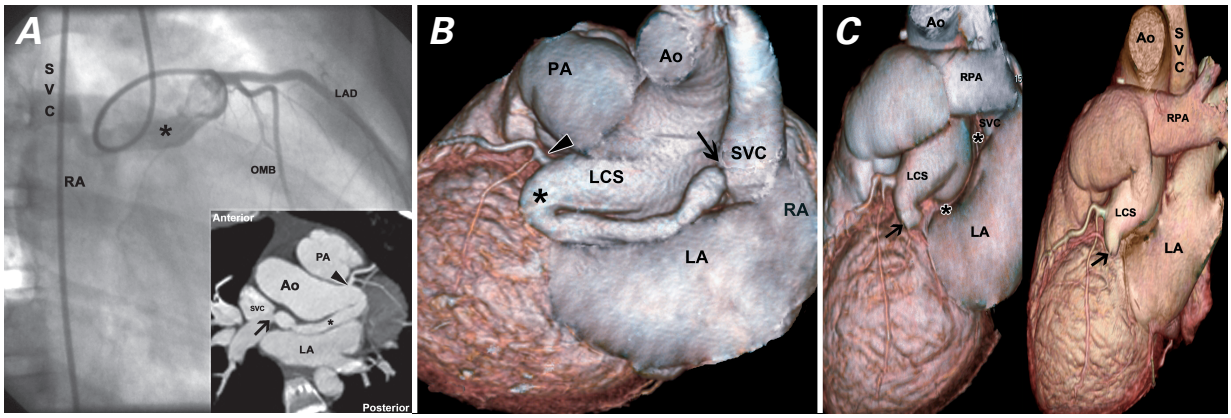

Fig. 1 *Patient 1. Aortic root injection. Angiography* ***A****) in right anterior oblique view and* ***A, inset****) computed tomography (CT) show the tunnel's origin (arrowhead) and termination (arrow).* ***B****) A CT angiographic reconstruction shows the close relationship between the tunnel and the left main coronary artery (arrowhead) in the left aortic sinus, the tunnel's retroaortic course, and its termination (arrow).* ***C****) Postoperative CT angiographic images after 3 months (left) and 18 months (right) show progressive obliteration of the tunnel (asterisks) and remodeling of the aortic root (arrows).*

** = tunnel; Ao = aorta; LA = left atrium; LAD = left anterior descending coronary artery; LCS = left coronary sinus; OMB = obtuse marginal branch; PA = main pulmonary artery; RA = right atrium; RPA = right pulmonary artery; SVC = superior vena cava*

closed with use of a GORE-TEX® patch (W.L. Gore & Associates; Flagstaff, Ariz), and the tunnel was ligated close to the greater curvature of the aorta. A patent cul-de-sac was left at the aortic wall. No residual shunt was seen on an end-operative transesophageal echocardiogram. At 1- and 3-month follow-up, 2D echocardiography revealed normal left ventricular and valve function. Follow-up CT angiograms at 3 months (Fig. 1C, left) and 18 months (Fig. 1C, right) showed progressive obliteration of the tunnel, remodeling of the left sinus, and normal coronary arteries.

Patient 2. In October 2008, an asymptomatic 24-year-old man was referred for evaluation of a continuous precordial murmur. Examination of the cardiovascular system revealed a hyperkinetic precordium and a continuous murmur at the right 2nd and 3rd intercostal spaces. Electrocardiography showed sinus rhythm, a normal axis, and right ventricular hypertrophy with a strain pattern. Chest radiography revealed a cardiothoracic ratio of 0.65. Echocardiography revealed a large anterior tunnel from the right aortic sinus that coursed retrocavally to the SVC–RA junction. Coronary angiography showed a Qp/Qs ratio of 1.6:1, aortic-root angiography revealed a faint filling of the right coronary artery, and CT angiograms yielded excellent definition of the anatomy (Figs. 2A and 2B).

Intraoperatively, the proximal portion of the tunnel, its relationship to the right coronary artery, and aneurysmal expansion at its termination were identified (Fig. 2C). With the patient under standard CPB, the tunnel was opened, its aortic end was oversewn, and the right coronary button was reimplanted into the aortic sinus. The RA opening was obliterated through direct suture approximation. The patient made an uneventful recovery and was doing well at his last follow-up examination, 10 months after surgery.

Discussion

Congenital aorta–RA tunnel, first described by Coto and colleagues in 1980,[1] is an extracardiac vascular communication between one of the aortic sinuses and the RA. An aneurysmal dilation of the sinus nodal artery has been proposed as the embryologic basis of this unusual lesion.[2] However, the origin of the tunnel from the noncoronary sinus cannot be explained by this theory.[1,3] Other explanations include an abnormal formation of the supravalvular ridge and persistent mesocardial cysts.[1]

A more likely cause is a congenital deficiency of the elastic lamina in the aortic media, which gradually enlarges under the influence of high aortic pressure to form an extracardiac tunnel. The preferential anterior or posterior course of the tunnel (from the right or left sinus of Valsalva, respectively) relates to direct anatomic proximity to the low-pressure RA.[4] The lesion results in an atrial-level left-to-right shunt.

The absence of myocardial branches differentiates aorta–RA tunnel from a coronary–cameral fistula. The tunnel's origin above the supra-aortic ridge differentiates it from a sinus of Valsalva aneurysm.[5] Patients with an aorta–RA tunnel may be asymptomatic, or they may present with exertional breathlessness, palpitations, or recurrent respiratory tract infections.[4] During periods of increased myocardial oxygen demand, a relatively greater fall in the resistance and subsequent dilation of the coronary arteries prevents coronary steal phenomenon through the tunnel.[6] The continued patency of the tunnel poses these risks: calcification of its

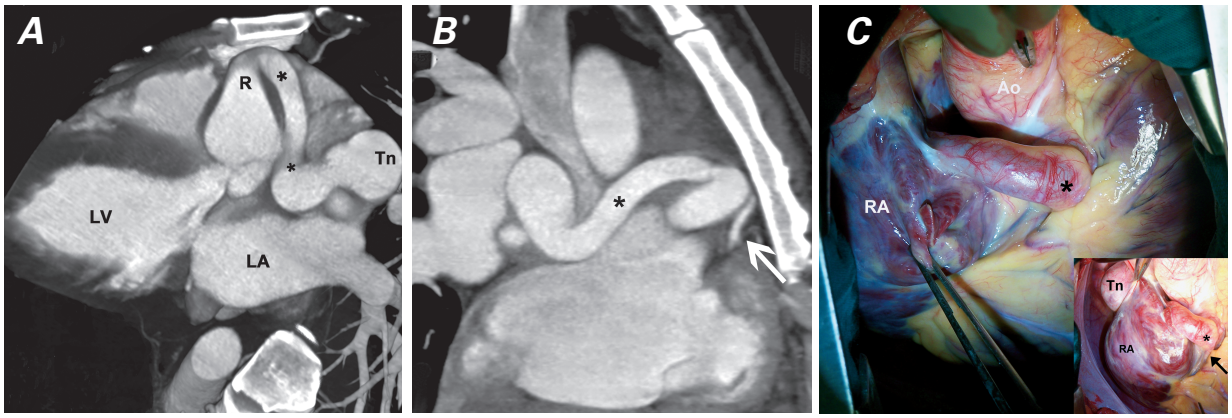

Fig. 2 *Patient 2. Computed tomographic angiograms show **A**) the origin (R), course (*), and termination (Tn) of the tunnel, and **B**) the course of the tunnel (*) and the origin of the right coronary artery (arrow) (sagittal view). **C**) Intraoperative photograph. Inset shows the right coronary artery (arrow).*

** = tunnel; Ao = aorta; LA = left atrium; LV = left ventricle; R = right aortic sinus; RA = right atrium; Tn = distal aneurysmal segment of the tunnel*

wall; aortic regurgitation[2]; biventricular volume overload or aneurysmal expansion[1,7]; or congestive heart failure, pulmonary vascular disease, infective endocarditis, and a higher mortality rate during surgery if the lesion is left uncorrected until a patient ages.[4,7]

Treatment options include catheter-based intervention, ligation under controlled hypotension, or repair with the patient under CPB.[4,8] External ligation of the tunnel close to the aorta should be performed only after accurate evaluation of the external anatomy and of the relationship between the coronary ostia and the orifice of the tunnel at the aortic end.[3] The aortic orifice may be left undisturbed in order to prevent inadvertent injury to the coronary ostia.[4,9] If the coronary artery arises from the tunnel, an alternative is to reimplant the artery as a button into the aortic sinus.[10]

Conclusion

Aorta–RA tunnel is a rare lesion of uncertain cause. It is often detected during the evaluation of a heart murmur in an asymptomatic patient. The lesion may respond to catheter-based therapy. If surgical repair is undertaken, either of 2 procedures—simple external ligation under controlled hypotension or closure of the RA orifice alone with the institution of CPB—may suffice, because a higher pressure in the aorta and the coronary arteries should prevent retrograde extension of thrombus from the tunnel. In our 1st patient, in view of the proximity of the tunnel's orifice to the left coronary ostium in the aortic root, we adopted the safer approach of external ligation, leaving a patent cul-de-sac at the aortic end. We considered the possibility of mural thrombosis, its extension or embolism into the coronary branches, and secondary infection in the remnant portion of the tunnel, and therefore placed the child under close clinical and radiologic follow-up. Postoperatively, she was clinically asymptomatic, and early and late follow-up imaging revealed normal coronary arteries, no aneurysmal changes, and progressive obliteration of the tunnel with remodeling of the aortic root.

Aorta–RA tunnel has been widely reported. We have discussed our experience in order to highlight the outcome on follow-up imaging (in patient 1) and to stress the importance of the identification and safe reimplantation of the right coronary artery (in patient 2).

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