AORTICO-RIGHT ATRIAL TUNNEL: Case of a Rare Congenital Communication

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Summary

An unusual case of aortico-right atrial tunnel is presented. The patient was referred to our institution for evaluation of a continuous heart murmur best heard along the right upper sternal border. Ascending aortography showed the tunnel taking its origin from the aortic root and entering the right atrium through a tortuous link. The patient underwent sugical repair because of a large left to right shunt. ***(Arch Turk Soc Cardiol 2003;31:298-302)***

Key words: *Aortico-right atrial tunnel, congenital anomaly, large left-to-right shunt*

Özet

Aorta-Sağ Atriyum Tüneli: Nadir Doğumsal Bağlantı

Nadir görülen aortik-sağ atriyal tünel olgusu sunulmaktadır. Olgu, sağ üst parasternal bölgede devamlı üfürüm nedeniyle merkezimize gönderildi. Assendan aortografi, aort kökünden kaynaklanan ve tortüyöz bir yapı ile sağ atriyuma giren tüneli gösterdi. Ciddi sol-sağ şant nedeniyle operasyon uygulandı. ***(Türk Kardiyol Dern Arş 2003;31:298-302)***

Anahtar kelimeler: *Aorta-sağ atriyum tüneli, doğumsal anomali, geniş sol sağ shunt*

The aortico-right atrial tunnel is a rare congenital anomaly first described by Otero Coto and his colleagues in 1980[1]. Anatomically, it is similar to aortico-left ventricular tunnels in that the aneurysmal communication originates from the aorta independent of the coronary arteries. In this pathology, there is a vascular link arising from the aortic root and terminating in the right atrium. We present a new patient with this unusual communication that has been successfully treated surgically. We compare our patient's characteristics with seven previously reported cases in the literature.

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REPORT of CASE

A 29-year-old man was referred to our institution for evaluation of a continuous grade 3/6 heart murmur best heard along the right upper sternal border. The electrocardiography showed right atrial enlargement. The chest x-ray revealed mildly increased pulmonary vascularity. Echocardiographic examination identified a large structure coursing from the right sinus of Valsalva toward the mildly dilated right atrium. A parasternal long and short axis views demonstrated a large structure in continuity with the right sinus of Valsalva (Fig. 1 and 2). An apical four chamber view showed a cross section of a vessel passing parallel to the atrioventricular groove in front of the right atrium, extending toward the junction of the superior vena cava with the right atrial roof. Doppler evaluation of this structure revealed continuous, turbulent flow in the right atrium.

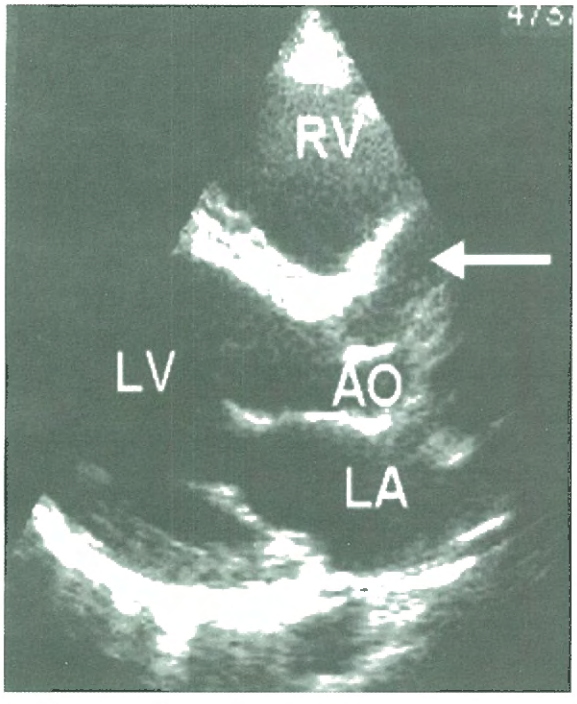

Figure 1: *In the parasternal long axis view, the large canal taking origin from the right sinus of Valsalve could be seen (arrow).*

AO:aort, LA: left atrium, LV: left ventricle, RV: right ventricle

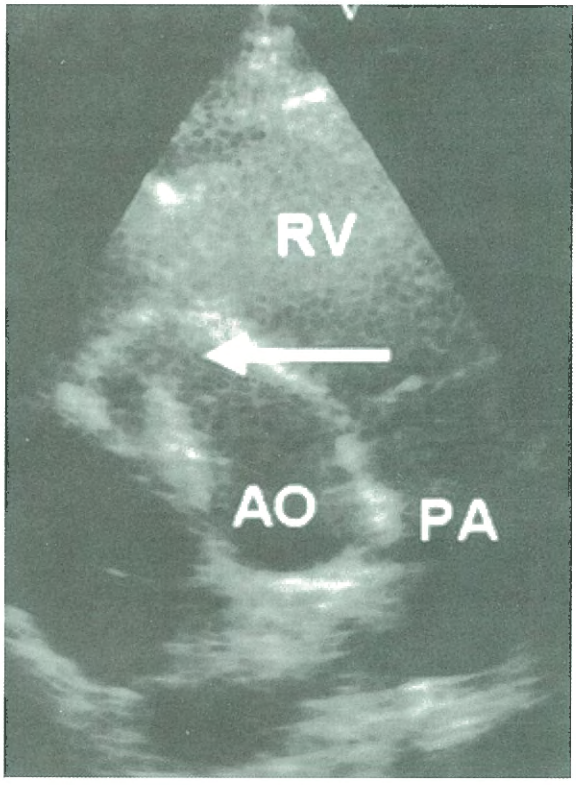

Figure 2: *In the parasternal short axis view, the large canal taking origin from the right sinus of Valsalve and its proximal course could be seen (arrow).*

AO:aort, RV: right ventricle, PA: pulmonary artery

Aortography identified the presence of a large tunnel originating from the right sinus of Valsalva, progressing in the right direction of the aorta, passing parallel to right atrioventricular groove and terminating in the roof of the RA. Injection of contrast material into the aortic root resulted in rapid filling of a large canal taking origin from the right sinus of Valsalva, right atrium, right ventricle and pulmonary artery (Fig. 3). The hemodynamic study revealed a set-up in oxygen saturation at right atrial level suggestive of a left-to-right shunt with a pulmonary-systemic blood flow ratio of 2.1: 1. Pulmonary artery pressure was measured 30/15 mmHg. Coronary angiography showed normal coronary arteries and no sinus node artery could be identified. The right coronary artery could be identified by a right Judkins catheter. An operation was planned because of a significant left-to-right shunt. At surgery was noticed that the right coronary

artery arose from the tunnel and its orifice was appoximately 2 cm from the tunnel's origin at the right sinus of Valsalva (Fig. 4). The tunnel was opened to the right atrium and the incision was then extended toward the right coronary ostium arising from the tunnel. The tunnel was completely dissected up to its right atrial origin and cut from the wall of the right atrium. The defect was then sutured with a running suture at the level of its origin in the right atrium to the ostium of the right coronary artery. Thereafter, the tunnel was horizontally incised with care taken to protect the right coronary ostia and its wall resected to reduce the lumen size. The remaining walls were sutured face to face. The control echocardiography findings were normal and the patient was asymptomatic in follow-up examinations.

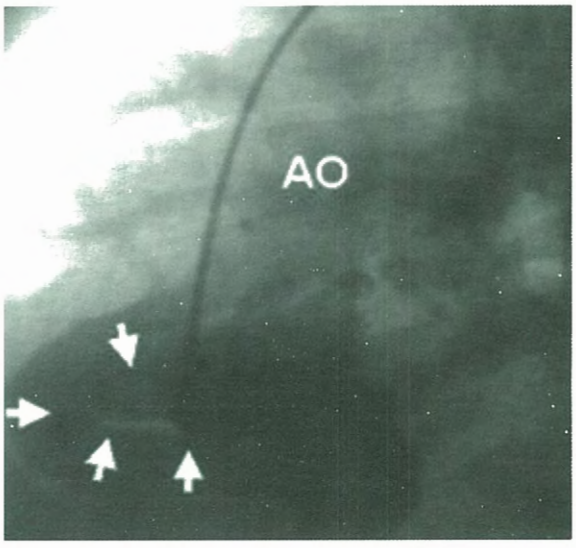

Figure 3: *Thoracic aortography in left oblique projection. Injection of contrast material into the aortic root resulted in rapid filling of a large canal taking origin from the right sinus of Valsalva (arrowheads).*
AO: aort

Table 1: *Characteristics of patients in the literature.*

Author	Year	Number of patients	Age and sex	Catheterization findings (Qp/Qs)	Morphologic characteristics of tunnel (site of origin and termination)	Coronary artery features
Otero et al.	1980	1	25-year-old man	Small	immediately above the noncoronary sinus-lateral aspect of the RA	Coronary arteries arose normally
Rosenberg et al.	1986	4	7-year-old girl	Small	above the left sinus of Valsalva-roof oft the right atrium	Coronary arteries arose normally
			6-month-old girl	1.7:1	above the left sinus of Valsalva-the superior aspect of the RA	LMCA arose from the mounth of the tunnel
			15-year-old boy	1.3:1	above the left sinus of Valsalva-the superior vena cava near its junction with the RA	Coronary arteries arose normally
			8-month-old male infant		above the left sinus of Valsalva-the superior vena cava near its junction with the RA	LMCA arose from the tunnel
Kalangos et al.	2000	2	18-year-old man	1.5:1	the left sinus of Valsalva-the roof of the RA	Coronary arteries arose normally
			7-year-old boy	1.3:1	the left sinus of Valsalva-the roof of the RA	Coronary arteries arose normally

RA: right atrium, LMCA: left main coronary artery

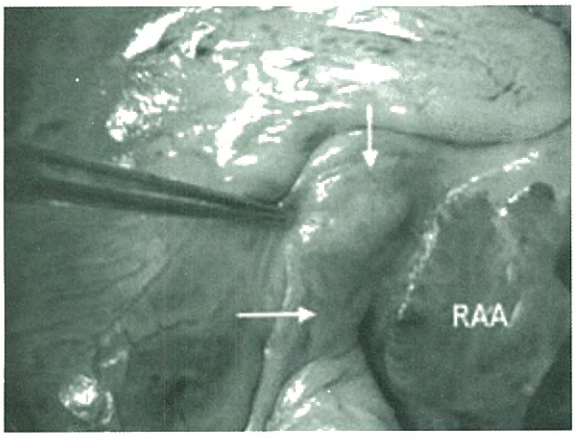

Fig. 4: Operative view showing the aortico-right atrial tunnel (arrows). RAA: right atrial appandix, AO: aort

DISCUSSION

Congenital aorta-cameral communications are rarely described in the literature. Although connection between aorta and left ventricle has received the most attention, there are only a few sporadic case reports describing the other aortico-cameral connections such as between aorta and right atrium, aorta and right ventricle, aorta and left atrium[2-5]. Literature concerning aortico-right atrial communications is limited to 7 patients (Table 1). Anatomic characteristics of these patients were different. The communication between the aortic root and the right atrium arose from above the noncoronary sinus in 1 patient , from above the left sinus of Valsalva in 4 patients, and from inside the left sinus in 2 patients, respectively[1,3,4]. In our case, it arose from above the right sinus of Valsalva, which has been described. In the literature, the communications were terminated in the lateral aspect of the right atrium, in the superior vena caval junction of the right atrium or in the roof of the right atrium as in our case. In some patients communication arose independently from the coronary artery, while in the others one of the coronary arteries originated from the communication. The distribution of both coronary arteries appeared normal in all patients and no sinus node artery could be identified in 4 patients. In our case, although the right coronary artery could be identified by a right judkins catheter in coronary angiography, it was observed to be originating from the tunnel. Authors have suggested some explanations on the tunnel's origin. Fistulous involvement of the sinus node artery suggested as an explanation for the origin of the tunnel because of its absence. But no sinus node dysfunction was seen in any patients, including our patient. Bharati at al.[5] reported an anomaly which they called aortico-right ventricular tunnel. The abnormal formation of the supravalvular ridge leading to weakness in the aortic wall was thought as a cause of communication in their report. Goor et al.[6] thought that the presence of mesocardial cysts found in various sites on the surface of the epimyocardium in the early stages of cardiogenesis might have caused the tunnel formation. Whether there is a link between these theories and the tunnel formation is unclear. The diagnosis of aortico-right atrial communications was revealed as a result of evaluation of a continuous heart murmur in essentially asymptomatic patients. The murmur heard at the right sternal border or in the left infraclavicular area should lead cardiologists to further investigations. Although two-dimensional echocardiography is a useful noninvasive diagnostic tool in patients with heart murmur, echocardiographic findings were not useful except for markedly dilated sinus of Valsalva. In our patient, a large structure arising from above the right sinus of Valsalva and terminating in the right atrium could be seen in various echocardiographic views. In addition, Doppler evaluation of this structure revealed a continuous, turbulent flow. Ascending aortography combined with selective coronary angiography and a hemodynamic study established the correct diagnosis.

The differential diagnosis should include a ruptured aneurysm of the sinus of Valsalva, a coronary arteriovenous fistula, a rupture of a

dissecting aneurysm of the ascending aorta into the right atrium[7], and a pseudoaneurysm of the right coronary artery followed by the formation of a fistula between the aneurysm and the right atrium[8]. We think that these diagnoses are not supported in the absence of predisposing factors and symptoms suggestive of a rupture. Due to the absence of small myocardial branches, coronary cameral fistula was not considered. In addition, the findings of surgical operation have provided more definitive information. The histological examination of tunnel material demonstrated that its tissue is similar to the aortic wall with an intimal thickness, medial degeneration and connective tissue proliferation. The need for operative closure in asymptomatic patients is not clear. In our case, a significant oximetric step-up which was greater than in previously reported cases at the atrial level indicated the necessity of surgical operation. The possible complications of the tunnel in unoperated patients include volume overload of both ventricle, bacterial endocarditis, aneurysm formation, and a spontaneous rupture. In conclusion, the aortico-right atrial tunnel should be considered in the differential diagnosis of continuous heart murmurs that tend to be heard at the right upper sternal border. Echocardiography and ascending aortography combined with coronary angiography, are the diagnostic tools which demonstrate its origin from the aortic root and its terminating site. Surgical closure is recommended in patients with a large left to right shunt in order to prevent aforementioned risks. Coil embolization could be thought of as a less invasive treatment strategy in patients whose coronary artery does not arise from the tunnel. We suggest that regular follow-up should be recommended especially in patients whose tunnel wall has not been totally excised, since the histologic characteristics of the wall may predispose patients to various complications.

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