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Aortico-cameral communication from right sinus of valsalva to right atrium

Sağ valsalva sinüsü ile sağ atriyum arasında aertico-kameral komünikasyon

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Aortico-right atrial tunnel (ARAT) is a rare congenital entity. We report a case of 57-year-old man who presented with unstable angina and who had a continuous murmur at the right sternal edge. Coronary angiograpy revealed triple vessel coronary artery disease and a separate anomalous structure arising from the right sinus of Valsalva. Digital substraction angiography was necessary to define the distal end of this connection which was the roof of the right atrium. Our patient is unique in that the tunnel 5 cm in lenght originated from the right sinus of Valsalva and entered the right atrium anteriorly having traversed the right atrioventricular groove anterior, totally separate to the right coronary artery. The management of these congenital aorto-atrial connections is discussed.

Key words: ***Aortico-Cameral-Fistula, Aorta-Atrium, Sinus of Valsalva***

Aortiko-sağ atriyal tünel nadir görülen konjenital anomalidir. Bu vaka takdiminde anstabil anjinası ve fizik muayenede sağ sternal bölgede devamlı üfürümü olan 57 yaşında erkek hasta sunulmaktadır. Koroner anjiyografide üç damar hastalığı ve sağ Valsalva sinüsünden ayrı olarak çıkan anormal bir yapı tespit edildi. Dijital substraksiyon anjiyografide bu yapının sağ atrium çatısıyla ilişkisi olduğu görüldü. Hastamızda, sağ koroner arterden tamamen ayrı olarak, sağ Valsalva sinüsünden başlayıp sağ atriyoventriküler oluğun ön kısmında sonlanan, 5 cm uzunluğunda bir tünel tespit edildi.Yazımızda bu tip doğumsal aorto-atriyal bağlantıların tedavisi tartışıldı.

Anahtar sözcükler: ***Aortiko-kameral fistül, Aorta-atrium, Valsalva sinüsü***

An abnormal connection between the aorta and cardiac chambers is a well recognised cardiac entity. Several types have been described, of which coronary-cameral fistulae or connections (congenital or acquired) are the most common (1). There have also been several reports of acquired aorta-cameral connections. Congenital aortico-cameral connections are much rarer and the majority are to the left ventricle (ALVT) (2;3). The aorticoright atrial tunnel (ARAT) is first described by Otero Coto and colleagues in 1980 (4). To date there have only been 13 cases reported of ARAT, most of which originated from the aortic left sinus of Valsalva whereas the tunnel in one case originated from the non-coronary sinus, in two cases from the right coronary sinus and in one case from the aortic isthmus (5-7). We report the third case of congenital ARAT arising from the right sinus of Valsalva.

A 57-year-old man was admitted with unstable angina and a four month history of exertional chest pain. He was a smoker with a positive family history of ischaemic heart disease. His past medical history was otherwise unremarkable. The chest roentgenogram revealed mild cardiomegaly and the ECG showed sinus rhythm with evidence of T-wave inversion in leads 1, aVL, V_2-V_6**.** On admission his cardiac enzymes were not elevated. He was managed conservatively and investigated.

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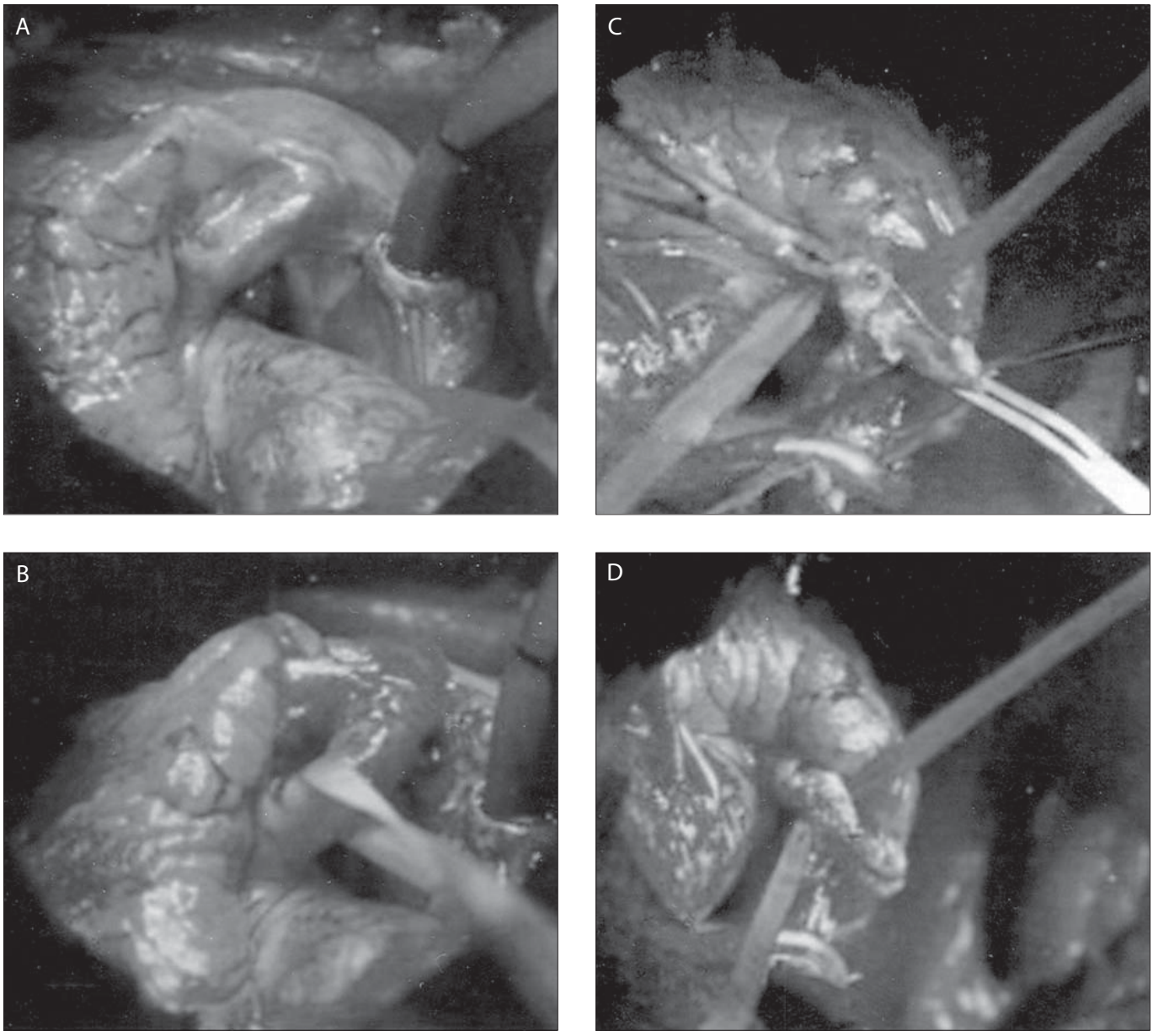

Figure 1. Intraoperative photograph showing aortico-right atrial tunnel (the patient's head is on the lower end); **(A)** A systolic thrill is verified by palpation, **(B)** blunt dissection is used for posterior tunnel attachments, avoiding injury to right coronary artery, and the tunnel was taped, **(C)** Clamps have been applied to the tunnel, with caution taken to monitor ECG changes before division, and completed division of the aortico-right atrial tunnel, **(D)** The ends of the tunnel are closed with 4-0 polypropylene suture. Ao=ascending aorta, RA=right atrium, RV=right ventricle.

Selective coronary angiography revealed a normal left main stem with significant triple vessel coronary artery disease and an anomalous tube like structure arising from the right sinus of Valsalva adjacent but totally separate from the right coronary artery and connecting the aorta to one of the cardiac chambers and gross macroscopic operative features confirmed these findings. (Fig.1). Transoesophageal echocardiography confirmed these findings. Digital substraction angiography (DSA) and CT scanning were, however, required to define the distal end of the connection as the roof of the right atrium. The ratio of pulmonary blood flow to systemic blood flow was measured as 1.9/1.

The patient underwent operation via a median sternotomy. A systolic thrill could be palpated over the base of aortico-right atrial connection. Cardiopulmonary bypass was established with moderate hypothermia (32 °C). After coronary artery bypass grafting to left anterior descending, intermediate, first obtuse marginal and right coronary arteries the anomalous structure was opened in the right atria-ventricular groove. The structure was described as 5-cm long fibrous tube anterior to the right coronary artery in the right atrioventricular groove with an external diameter of 1.5-cm. The tube was divided between vascular clamps and the cut ends over sewn. The patient was

Table 1. Reports on Surgically Treated ARAT in Literature

First Author, publication year	*Gender, age*	*Aortic Origin of ARAT*	*Symptoms*	*Associated anomalies*
Tanaka et al. 2005 (10)	Female, infant	?	Congestive heart failure	Aortic atresia, right-sided atrioventricular valve atresia, ventricular septal defect, atrial septal defect, and persistent ductus arteriosus
Kursaklioglu et al. 2005 (5)	Male, 27 yo.	Left sinus of Valsalva,	Asymptomatic	No sinus node artery
Turkay et al. 2003 (6)	Male, 29 yo.	Right sinus of Valsalva	Palpitation and effort dyspnea	None
Elwatidy et al. 2003 (7)	Female, 3 yo	Aortic isthmus	Asymptomatic	None
Tsai et al. 2002 (11)	Female, 2 yo.	Left sinus Valsalva	Asymptomatic	Marfan's syndrome
Kalangos et al. 2000 (9)	Male, 18 yo Male, 7 yo	Left sinus Valsalva Left sinus Valsalva	Asymptomatic Asymptomatic	Moderate aortic insufficiency, no sinus node artery No sinus node artery
Danilowicz et al. 1989 (12)	Female, Newborn	Right sinus of Valsalva	Congestive heart failure	Secundum atrial septal defect
Rosenberg et al. 1986 (8)	Female, 7 yo Female, 6 month old Male, 15 yo Male, 8 month old	Left sinus of Valsalva Left sinus of Valsalva Left sinus of Valsalva Left sinus of Valsalva	Asymptomatic Asymptomatic Asymptomatic Asymptomatic	None Left ventricular hypertrophy None Left ventricular hypertrophy
Otero Coto et al. 1980 (4)	Male, 25 yo	Noncoronary Sinus	Asymptomatic	Absence of the right superior vena cava with a large persistent left superior vena cava draining to the coronary sinus

weaned off bypass without difficulty. Apart from a temporary pacing requirement for nodal rhythm for 24 hours he made an uneventful recovery. Two years after the operation the patient remains asymptomatic.

Comment

Congenital aortico-cameral connections are extremely rare, but have been reported connecting the aorta to all four cardiac chambers. The majority of these are aortico-left ventricular connections which have been extensively reviewed and classified (3). Congenital ARAT is however, an extremely rare entity, and only 13 cases have been previously reported in the English literature (Table 1). Mostly arise from the left sinus of Valsalva. This case is unique in that the origin of the aortico-right atrial connection was the right sinus of Valsalva, adjacent but separate to the right coronary ostium and the patient presented with unstable angina. Furthermore, our case is the oldest patient in the literature presented with ARAT and surgically treated.

These connections can be difficult to define accurately as was experienced in this case, where conventional angiography and echocardiography were inadequate. CT scanning and DSA did, however, illustrate the value of these imaging techniques in demonstrating abnormal cardiac structures.

Differential diagnoses included ruptured aneurysm of the sinus of Valsalva, coronary arteriovenous fistula, rupture of a dissecting aneurysm of the ascending aorta into the right atrium, and pseudoaneurysm of the right coronary artery followed by formation of a fistula between the aneurysm and the right atrium. There was no history of trauma in our case and no evidence of acquired aortic disease. We conclude therefore, that this aortico-right atrial connection was a congenital structure. All previous congenital connections of this type have been described in the pediatric population. Our patient is unique in having such a congenital structure diagnosed at the time of presentation with a separate acquired cardiac condition. Indeed the histological findings confirm those described by Rosenberg et al. of a deficiency in smooth muscle, but abundance of elastic lamellae (8). The precise pathogenesis of this congenital anomaly is however unknown; persistence of mesocardial cysts or embryonic rests of the fifth aortic arch in the early stages of cardiogenesis, abnormal formation of supravalvular ridge producing aneurysmal dilatation of the primitive aorta, intrauterine rupture of a sinus of Valsalva aneurysm are possible explanations (4,8).

Previous reports have suggested that patients with aortico-right atrial tunnel may present with continuous heart murmur, mild cardiomegaly or clinical evidence of ventricular overload due to left-to-right shunt. Angina and myocardial infarction in contrast have not been reported secondary to aortico-right atrial communications. The ischemic symptoms in our patient could theoretically have been due to coronary artery steal, but in reality probably was due to the proximal stenosis of left anterior descending artery.

Of the congenital aortico-cameral connections, at least 37 cases have been reported of aortico-left ventricular tunnel (ALVT) (3). Surgery is clearly indicated in this condition to prevent progressive aortic incompetence and cardiac failure. Little, is however, known of the natural history of untreated aortico-right atrial connections. Congenital ARAT is also uncommon except for complications with a coronary artery fistula. We, however, assume that this communication was not an equivalent of a coronary artery fistula, since the coronary arteries in our case had normal origin, distribution and no dilatation. In general, surgical correction is recommended to avoid the long term effects of left-to-right shunting thus preventing left ventricular impairment and improving life expectancy. Furthermore because of possible complications such as bacterial endocarditis, aneurysm formation, or spontaneous rupture, the closure of ARAT is recommended (9). However, it should be kept in mind during surgery that the "tunnel" can also give origin to coronary arteries. Definitive surgical correction has been shown to be effective and safe, with good long term results.

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