

A newborn with Aortico-left Ventricular Tunnel mimicking Sinus of Valsalva Aneurysm

SENAY AKBAY¹, Filiz EKICI², FIRAT KARDELEN³, MUHAMMET BULUT⁴, Zeynep Mutlu¹, and Gökhan ARSLAN⁴

¹Akdeniz University Faculty of Medicine

²Akdeniz Universitesi

³AKDENİZ UNIVERSITY MEDICAL SCHOOL

⁴Akdeniz University Faculty of Medicine, Antalya

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Abstract

Aortico-left ventricular tunnel (ALVT) is a rare congenital cardiac anomaly and constitutes of less than 0.1% of all congenital cardiac defects (1). ALVT is described as an abnormal connection between the ascending aorta and the left ventricle which originates commonly above the right sinus of valsalva. Most patients are diagnosed with an ALVT during early infancy (2). Although transthoracic echocardiography (TTE) is more effective in diagnosis of ALVT, misdiagnosis rate was 17.1% (3). Sinus of valsalva aneurysm (SVA) is frequently confused with ALVT (3). We report a term female newborn with SVA in echocardiographic examination but in surgery she was diagnosed with ALVT.

Title

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Authors

Dr. Şenay Akbay, Fellow of pediatric cardiology, Department of Pediatric Cardiology 1. Akdeniz University Faculty of Medicine, Antalya

Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070,Kampüs, Konyaaltı/ANTALYA /TURKEY

E- mail: senayakbay@gmail.comTel: +90 242 2496543

Fax: +90 242 347 23 30 GSM: +90 (546) 407 27 37

Prof. Dr. Filiz Ekici, Department of Pediatric Cardiology 1. Akdeniz University Faculty of Medicine, Antalya

Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070, Kampüs, Konyaaltı/ANTALYA / TURKEY

E- mail: ekicifiliz@gmail.comTel: +90 242 2496543

Fax: +90 242 347 23 30 GSM: +90 (505) 466 36 20

Prof. Dr. Fırat Kardelen, Department of Pediatric Cardiology 1. Akdeniz University Faculty of Medicine, Antalya

Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070, Kampüs, Konyaaltı/ANTALYA/ TURKEY

E- mail:fkardelen@akdeniz.edu.trTel: +90 242 2496530

Fax: +90 242 347 23 30 GSM: +90 (505) 4170650

Dr. Muhammet Bulut, Fellow of pediatric cardiology, Department of Pediatric Cardiology 1. Akdeniz University Faculty of Medicine, Antalya

Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070, Kampüs, Konyaaltı/ANTALYA

E- mail: dr.mhbulut@gmail.com Tel: +90 242 2496543

Fax: +90 242 347 23 30 GSM: +90 (553) 2446546

Dr. Zeynep Çağla Mutlu, Fellow of pediatric cardiology, Department of Pediatric Cardiology 1. Akdeniz University Faculty of Medicine, Antalya

Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070, Kampüs, Konyaaltı/ANTALYA Tel:

E- mail: z.caglamutlu@gmail.com Tel: +90 242 2496546

Fax: +90 242 347 23 30 GSM: +90 (505) 7590022

Prof. Dr. Gökhan Arslan, Department of Radiology 2. Akdeniz University Faculty of Medicine,

Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070, Kampüs, Konyaaltı/ANTALYA/ TURKEY. E-mail: dr.gokhanarslan07@gmail.com Tel: none Fax: none GSM: +90 (532) 7228491

Corresponding author; Dr. Filiz Ekici

Department of Pediatric Cardiology 1. Akdeniz University Faculty of Medicine. Antalya. Address: Pınarbaşı Mahallesi Dumlupınar Bulvarı 07070, Kampüs, Konyaaltı/ANTALYA/ TURKEY E-mail: ekicifiliz@gmail.com Tel: +90 242 2496543

Fax: +90 242 347 23 30 GSM: +90 (505) 466 36 20

Abstract

Aortico-left ventricular tunnel (ALVT) is a rare congenital cardiac anomaly and constitutes of less than 0.1% of all congenital cardiac defects (1). ALVT is described as an abnormal connection between the ascending aorta and the left ventricle which originates commonly above the right sinus of valsalva. Most patients are diagnosed with an ALVT during early infancy (2). Although transthoracic echocardiography (TTE) is more effective in diagnosis of ALVT, misdiagnosis rate was 17.1% (3). Sinus of valsalva aneurysm (SVA) is frequently confused with ALVT (3). We report a term female newborn with SVA in echocardiographic examination but in surgery she was diagnosed with ALVT.

Case

A two day-old term female baby was admitted our unit due to history of ventricular septal defect (VSD) determined in fetal echocardiography at the 32. gestational week. She had no complaints and her physical examination was normal after birth except 3/6 systolodiastolic murmur along the left sternal border. In transthoracic echocardiography (TTE), we detected a giant aneurysm located anterior of the aortic ring with a diameter of 11,5x12,7 mm and a small secundum atrial septal defect. Parasternal long axis and five-chambers views showed that the aneurysmatic sac was related with aorta and left ventricle (Figure-1A, B). In diastol period of cardiac cycle, there were two shunt flows, one was seen through the aortic root into the aneurysmatic sac (velocity:3,4 m/sn) and at the same time shunt flow from aneurysmatic sac to the left ventricle (velocity:2,3 m/sn) (Figure-2, Video-1). At subcostal examination, color flow doppler echocardiography showed the entrance of the aneurysm to left ventricle. The aneurysm also occupied the right ventricular outflow tract and caused mild obstruction (Video-2). Computed tomography showed the giant extracardiac aneurysmatic sac between aortic root and truncus pulmonalis and a suspicion of SVA (Figure-3). The patient has been referred to the another cardiac center for surgery. The surgery was confirmed the diagnosis of ALVT.

Differential diagnosis of ALVT included SVA, VSD with aortic malformations, aortic valve insufficiency, four-leaflet aortic valve, aortic valve prolapse, infective endocarditis. It is difficult to differentiate SVA from ALVT. SVA usually originates from below the level of sinotubular junction of aorta and forms as a pocket pouch or petals. During the diastole period, increased arterial blood volume in the left ventricle expands the diameter of sinus (2). ALVT is often originates from the tubular aorta, above the coronary artery ostia rather than aortic valve sinus (2). Hovaguimian et al.(4) defined an anatomic classification of aorto-ventricular tunnels; Type 1: Slit-like opening at the aortic end with no valve distortion, Type 2: Large extracardiac aneurysm, Type 3: Intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow tract obstruction, Type 4: Combination of Types 2 and 3. Our case was compatible with type 4.

Although TTE is gold standard for definition of SVA and ALVT, the discrimination of these two entities can be difficult in some cases. In our case, also CT couldn't help differential diagnosis and certain diagnosis was determined by surgery.

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