

Left aorto-ventricular tunnel: A differential diagnosis to aortic regurgitation

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SUMMARY

Left aorto-ventricular tunnel is a rare congenital heart disorder. A then 19-year-old young lady was referred to our clinic for chronic rheumatic heart disease with aortic regurgitation. However, because of an almost continuous murmur, and an unusual transthoracic echo, she underwent various investigations, finally revealing a rare type 2 left aorto-ventricular tunnel. She had been variously diagnosed as chronic rheumatic heart disease with aortic regurgitation, bicuspid aortic valve with aortic regurgitation, and then possibly coronary arterial (cameral) fistula (based on 2D transthoracic echo) and finally left aorto-ventricular tunnel. This is the first case described in South East Asia who is asymptomatic, apart from a murmur, and her long journey to this final diagnosis illustrates the need to “chase shadows” on echoes, to come to a proper conclusion for the patient.

INTRODUCTION

Aorto-ventricular tunnel (AVT) is a rare congenital heart disorder. It is an anomalous tract that connects the ascending aorta usually just above the sinotubular junction to either the right or left ventricle, with the left being more common. This is different from a fistula which forms following the rupture of a dilated sinus of Valsalva and hence below the level of the coronary ostia.¹ This anomalous tract has normal aortic vessel like media structurally¹ and most likely arises because of an abnormal separation of the great vessels and possibly valve leaflets (conotruncal region).² There are cases reported 2 where there are even rudimentary valve leaflets in these tunnels, hence making it likely that this is at least one of the possible ways that this rare anomaly arises.

CASE REPORT

Miss N, then a 19-year-old lady was referred to us from our adult cardiology colleagues with a diagnosis of chronic rheumatic heart disease with aortic regurgitation (AR). This was because of a murmur picked up during a routine school examination in her secondary school years. During clinical examination, we noted the presence of an almost continuous murmur. Our initial transthoracic echo (TTE) performed by one of us, detailed a bicuspid aortic valve (BAV) with an aberrant jet that did not look like AR. At the time, we thought of possibilities of coronary artery (cameral) fistula (CAF) with exit to the left ventricle or unlikely a left aortic-

ventricle tunnel (LAVT). We referred her to a large tertiary centre in Malaysia, but she was told that the diagnosis was BAV with AR, after a TTE performed there. She was then referred back to us for continuation of care. However, because of our initial echoes, we managed to persuade her family to agree for further investigations with the view of percutaneous closure from interventional catheterization. A repeat TTE performed, by another of the authors, was unusual (Fig. 1a) and clinically the agreed upon auscultation was that the murmur was continuous. The transoesophageal echo (TEE) showed clearly a LAVT (Fig. 1b). An angiogram performed in the right oblique view at 30° also revealed LAVT (Fig 2a). Because of that, the procedure was abandoned as none among the authors had experience with AVT, nor had read on possible percutaneous closure of LAVT. A CT angiogram scan (with contrast) was arranged, in the likelihood that the patient required surgery, and confirmed our diagnosis (Fig. 2b and c).

DISCUSSION

AVT is a rare congenital cardiac disorder that was first described in 1963 by Levy and colleagues.¹ It describes the lesion of a channel that connects the aorta (usually above the aortic sinuses) to the right, or more commonly left ventricle.³ A classification scheme based on the local anatomic findings of the AVT was proposed in 1988 by Hovaguiman and colleagues⁴ with:

Type I: A slit like aortic orifice without valvular distortion.

Type II: A larger, oval shaped aortic orifice with an extracardiac aneurysmal tunnel.

Type III: An oval aortic orifice with a septal (intracardiac) aneurysmal tunnel.

Type IV: A combination of type II and III.

This rare condition, has had various incidences reported ranging from 0.03% to 0.46% of all congenital heart defects.⁵ The clinical presentations can be varied, ranging from usually heart failure in infancy to the mildly symptomatic case. In our patient apart from the murmur, she was not in heart failure and was asymptomatic, even beyond her second decade of life. And it had been variously diagnosed as AR due to rheumatic fever or BAV with AR, and CAF. We believe this is the first reported case in South East Asia, and although rare, the abnormal colour Doppler image should pique interest in the possibility of other conditions. In reality, one of

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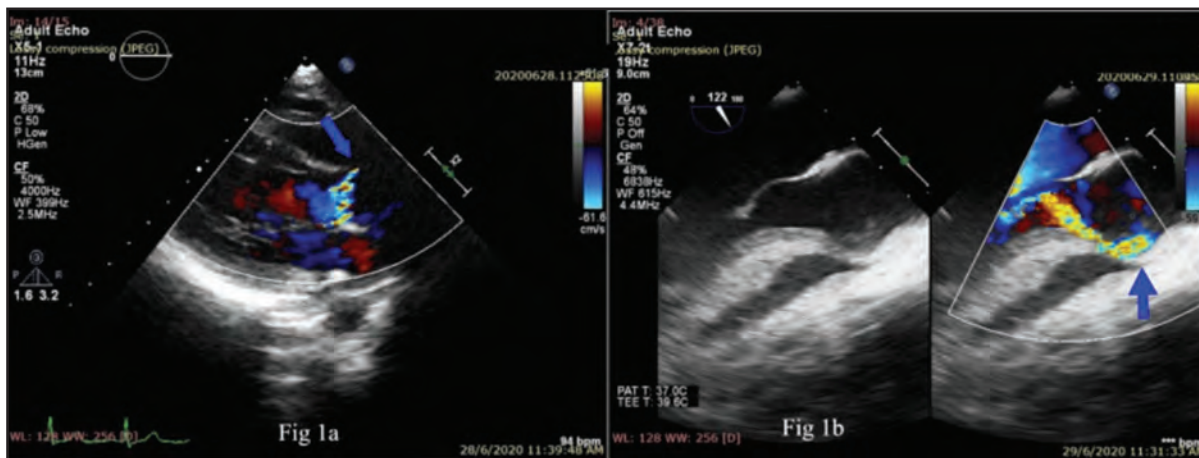


Fig. 1: Echocardiogram of the patient on long axis view (1a) showing skewed jet from patient with continuous murmur variously diagnosed as BAV and AR, AR secondary to rheumatic fever. Transoesophageal echo revealing left AVT (1b), which can be seen especially with colour Doppler, appearing above the coronary sinus and then coursing beside it before entering the left ventricle just below the aortic valve (arrow). AVT=aorto-ventricular tunnel, AR=aortic regurgitation, BAV=bicuspid aortic valve.

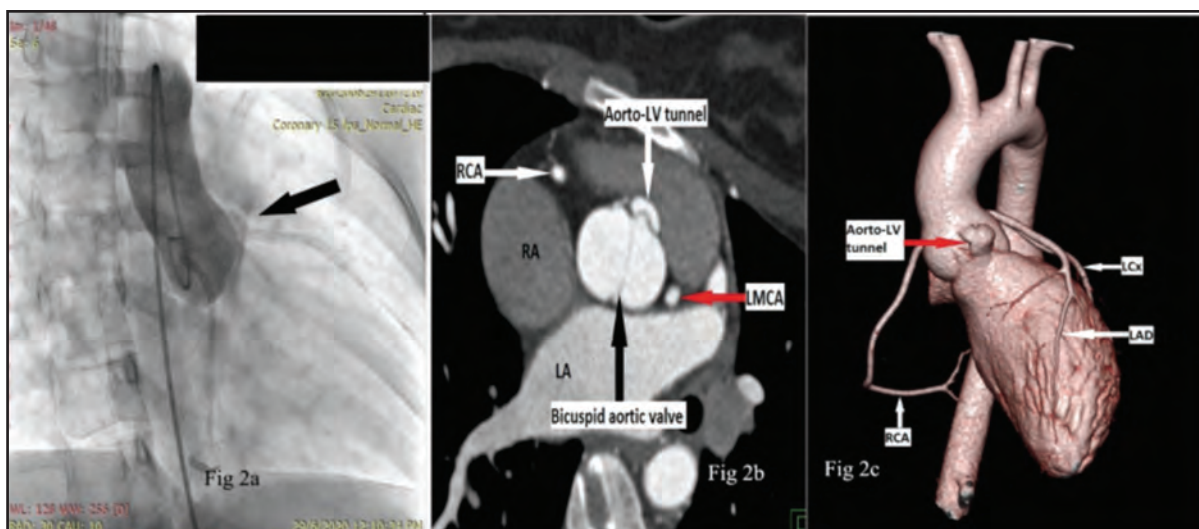


Fig. 2: An angiogram of the patient from the RAO 30o view (Fig 2a) revealing the left AVT (arrow). The top right corner was blacked out to remove the patient's details. AVT=aorto-ventricular tunnel. RAO (right anterior oblique) CT angiogram of the patient at the level of the aortic valve (Fig 2b). Left AVT is labelled (fine arrow). Reconstructed 3D CT image (Fig 2c) showing mainly the left ventricle, aorta and coronary vessels and left AVT (labelled). LA=left atrium, LAD=left anterior descending artery, LCX=left circumflex artery, LMCA=left main coronary artery, RA=right atrium, RCA=right coronary artery.

us tried tracking that jet and traced it back to a vascular-like structure, hence CAF was considered, and also LAVT. Because we thought the large centre was better equipped to deal with such situations, hence, the patient was initially referred there. After her return, we managed to persuade her and her family of the need for a proper diagnosis, hence we proceeded with TEE and also catheterization.

We did not proceed with any intervention, due to our lack of experience in LAVT, and upon perusing the literature, most discussed surgery and the importance of closing both ends of the track. Percutaneous closure results were also discouraging compared to surgery.⁶ This is understandable, considering that the echo and angiography are two dimensional images displaying a rather small track above the aortic sinus, and traversing to a region below the aortic valve. However, the 3D reconstruction on CT (Fig 2 b and c)

showed that although narrow in 1 dimension, the LAVT was wide in another with a curved extramural course (Type II), hence making percutaneous intervention difficult with many devices, unless one was specifically designed for it. Little to nothing is known of the natural history of asymptomatic cases, with one case of “spontaneous closure”⁷ and some reporting symptomatic individuals presenting at 45 years of age.⁶ But recommendations for LAVT are for closure due to the possible progression of heart failure and also possible progressive aortic regurgitation and dilation of the aorta.^{6,7}

CONCLUSION

LAVT should be considered as part of the differential diagnosis to AR, when there is an abnormal colour Doppler image, and when clinically, an almost continuous murmur can be heard.

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