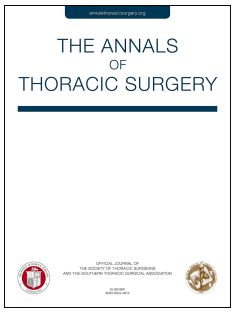

Aberrant Aorto Pulmonary Venous Fistula – an Intra-operative Surprise

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Running Head: ABERRANT AORTO PULMONARY VENOUS FISTULA

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ABSTRACT

Anisolated systemic artery to pulmonary vein arterio-venous fistula is a rare clinical entity. We report a 20 year old ladydiagnosed with myxomatous mitral valve prolapse with severe mitral regurgitation and planned for mitral valve repair. An aberrant aorto pulmonary venous fistula was suspected intraoperatively due to flooding of the left atrium with blood from the left inferior pulmonary vein on cardio pulmonary bypass. The mitral valve was repaired successfully. A post-operative Computed Tomographic angiogram revealed an anomalous fistula between the descending thoracic aorta and left inferior pulmonary vein. She underwent successful percutaneous device closure of the fistula.

Abstract word count: 99

Keywords: aorto-pulmonary venous fistula, intraoperative, mitral valve repair

Arterio-venous fistulae characterized by an aberrant systemic artery arising from the descending aorta inserting into one of the pulmonary veins are an extremely rare anomaly of cardiovascular anatomy. It causes a left to left shunt without lung sequestration. It may be congenital or acquired. It is associated with a continuous murmur of atypical location, bounding peripheral pulses and left ventricular enlargement. The lack of increased pulmonary flow and absence of pulmonary parenchymal or bronchial abnormalities differentiate it from other arterio-venous fistula/malformation of lungs. [1, 2].

We report a young lady who presented to us with dyspnea and was diagnosed pre-operatively with myxomatous mitral valve prolapse with severe mitral regurgitation. However, an intra-operative surprise awaited us during the mitral valve repair leading to a post-operative diagnosis of systemic to pulmonary vein fistula by Computed Tomographic angiogram and its eventual percutaneous closure with embolization.

A 20 year old lady presented to us with history of dyspnea on exertion which was of insidious onset and gradually progression [New York Heart Association (NYHA) Class II to Class III over 2 year duration]. It was associated with generalised fatigue. She had no co-morbidities or significant family history. On clinical examination, there were no signs of right heart failure or pulmonary hypertension. She had cardiomegaly (apical impulse at the 6thintercostal space 2 cm lateral to the mid clavicular line) and a grade 4/6 apical pan systolic murmur with thrill with a functional diastolic murmur.

Echocardiography demonstrated severe mitral regurgitation characterised by a central jet with postero-lateral deflection. The mitral annulus was dilated (45mm). The left atrium was large (5.7x5.1cm). The anterior mitral leaflet was found to be lengthy, redundant and prolapsing. The other valves were normal and the left ventricle was mildly dilated with no dysfunction. A diagnosis of myxomatous severe mitral regurgitation was made.

She was offered mitral valve repair through a lower midline hemi-sternotomy. Under cardio-pulmonary bypass, the heart was cross clamped and ante grade Del Nido cardioplegia was delivered. The heart arrested promptly and the mitral valve was approached trans-septally. But on incising the inter-atrial septum, the left atrium was found to be flooding with blood. On inspection, the blood was found to be filling exclusively from the left inferior pulmonary vein, thereby excluding a patent ductus arteriosus. A cardiotomy suction was placed in the pulmonary vein, which drained blood amounting to 20-30% of the total cardiac output. Patient was immediately cooled to 25^0C. An aberrant systemic to pulmonary vein fistula was suspected. The mitral valve was assessed and was found to have annular dilation and essentially normal leaflets. A mitral annuloplasty was done with 32mm Medtronic 3D profile™ ring (Medtronic plc, Minneapolis, Minnesota). The surgery was completed with intermittent periods of low flows. Intra-operative saline insufflation test showed no leak. Left ventricular (via right superior pulmonary vein and left atrium) and pulmonary artery vents were placed to prevent flooding and distension of the left ventricle. The cardiac chambers were closed. Continuous warm, normokalemic, retrograde cardioplegia was administered prior to closure of the inter-atrial septum to ensure that the heart was beating before removal of the cross clamp. The post-cardio-pulmonary bypass transesophageal echocardiogram showed no mitral regurgitation and laminar flow across the mitral valve. Interrogation of the left lower pulmonary vein on echocardiogram was inconclusive. The patient tolerated the surgery well and had an uneventful recovery

The patient was evaluated post-operatively with a computed tomographic angiogram, which revealed a long (16x30mm), tortuous fistulous tract arising from the left lateral aspect of the descending thoracic aorta at the level of D8 vertebral body. The neck of the tract measured 11.2mm. Distally the tract inserted unto the dilated left inferior pulmonary vein in close proximity to the lung parenchyma (Figure 1, 2). The patient underwent percutaneous closure of the aorto-pulmonary vein fistula with a 5/7 KONAR-MF™ device (Lifetech Scientific, Guangzhou, China) (Figure3)

The post-operative echocardiogram showed no mitral regurgitation and no residual flow in the aberrant fistulous tract. She was discharged in NYHA functional Class I and is on regular follow-up.

COMMENT

A congenital systemic artery to pulmonary vein fistula is an uncommon clinical entity diagnosed in infants and children with or without other associated congenital cardiac anomalies. The diagnosis of an isolated congenital fistula of this nature in adults is exceedingly rare. The presence of an aberrant systemic to pulmonary vein fistula is suspected clinically by features of the hemodynamically significant left to left shunt. In literature, the fistula has been diagnosed by Doppler or contrast echocardiography and angiography by cardiac catheterisation or computed tomography.[3-5]

Our patient presented with predominant clinical and echocardiographic features of severe mitral regurgitation. On retrospective analysis, the continuous murmur of the arterio-venous malformation could have been masked by the loud pan systolic murmur and the functional diastolic murmur of the mitral regurgitation. She did not have bounding peripheral pulses, probably due to the low forward flow due to the severe MR. This variety of arterio-venous fistula does not cause increased pulmonary arterial flow. The absence of pulmonary parenchymal or bronchial abnormalities, including lung sequestration makes them difficult to suspect pre-operatively. The patient can be asymptomatic clinically unless the vascular malformation has led to vascular compression or congestive cardiac failure[2]. The pre-operative symptoms reported by our patient were explained by the diagnosis of mitral regurgitation and hence an active search for such a malformation with a pre-operative angiography was not carried out.

The left atrium being flooded intra-operatively on cardiopulmonary bypass in a cross clamped heart led us to look for a cause for the left to left shunt. The high flow steal of systemic circulation to the vent via the fistula necessitated hypothermia to prevent end organ damage and intermittent low flows to permit visibility. The adequacy of systemic perfusion was monitored with venous saturation, lactates and urine output. Additional cerebral monitoring like near-infrared spectroscopy (NIRS) would be desirable. Left ventricular distension was prevented by adequate venting and rapid institution of cardiac activity. Intra-operative assessment of the mitral leaflets revealed annular

dilation. This is consistent with functional mitral regurgitation caused by left atrial volume overload and enlargement, probably caused by the increased flow from the left inferior pulmonary vein.

In retrospect we are now entertaining the possibility that a pre-existing mitral regurgitation could have been significantly aggravated by the fistula. The severity of the mitral regurgitation could have in turn led to the camouflaging of the clinical features of the vascular malformation. The unanticipated intra-operative diagnosis of the congenital systemic artery to pulmonary vein fistula and it's hereunto unreported association with functional severe mitral regurgitation makes the above reported case scenario unique. An untoward delay in intra-operative identification and management of the fistula could have resulted in severe ventricular distension and myocardial damage.

REFERENCES

1. Scott LP, 3rd, Perry LW. Systemic artery–pulmonary vein fistulas, congenital and acquired left to left shunt. *Am J Cardiol*. 1968;23:872–876.
2. Jariwala P, Ramesh G, Sarat Chandra K. Congenital anomalous/aberrant systemic artery to pulmonary venous fistula: closure with vascular plugs & coil embolization. *Indian Heart J.* 2014;66:95–103.
3. Robida A, Eltohami EA, Chaikhouni A. Aberrant systemic artery-pulmonary venous fistula: diagnosis with Doppler imaging. *Int J Cardiol*. 1992 Jun;35:407–411.
4. Wolf WJ, Casta a, Swischuk L. Aberrant systemic artery-pulmonary vein fistula: detection of an occult lesion by contrast echocardiography. *Am Heart J*. 1985 Aug;110:480–482.
5. Shebani SO, Khan MD, Tofeig MA. A congenital fistula between the descending aorta and the right pulmonary vein in a neonate presenting with heart failure. *Cardiol Young*. 2007 Oct;17:563–564.

FIGURE LEGENDS

Figure 1: Contrast enhanced Computed Tomographic image of the aberrant arterio-venous fistula arising from the lateral wall of the descending thoracic aorta and inserting into the left inferior pulmonary vein.

Figure 2: 3D reconstruction of the Computed Tomographic image of the aorto-pulmonary vein fistula.

Figure 3: Fluoroscopic image of the closure of the aorto-pulmonary vein fistula with a device.