AN AORTOPULMONARY TUNNEL AS AN EXTRA-CARDIAC SYSTEMIC TO PULMONARY SHUNT

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ABSTRACT

Aortopulmonary window, one of the rarest congenital heart diseases, causes unrestricted left-to-right shunt and may lead to congestive heart failure, pulmonary vascular obstructive disease, and low cardiac output. Echocardiogram and CT angiography are important non-invasive diagnostic tools. Surgical closure is indicated in all patients diagnosed with APW and is usually performed at the time of diagnosis to reduce the risk of developing early pulmonary vascular disease. We report on a term baby girl with variant of aortopulmonary window which manifest as a tunnel on CT angiography.

Keywords: Aortopulmonary window; Aortopulmonary tunnel; Sytemic-pulmonary shunt

INTRODUCTION

Aortopulmonary window (APW) is one of the rarest congenital heart diseases with systemic to pulmonary shunt. This case report aims to highlight a variant of APW which has tubular communication rather than the conventional defect between the ascending aorta to the main pulmonary artery. To date, there has been no consensus in naming such anomaly.

CASE PRESENTATION

A term baby girl was delivered via emergency Caesarean-section due to foetal distress. She was treated for congenital pneumonia and physiological jaundice. Following an incidental detection of heart murmur at day 4 of life, she was then referred to Paediatric Cardiology Unit. During a follow-up visit at day 22 of life, she was mildly tachypnoeic with mild subcostal recessions. Respiratory rate was 44 breaths per minute, with oxygen saturation of 98% on room air. An ejection systolic murmur at left sternal edge was noted. She had lost weight at a rate of approximately 300 g/day at day 22 of life. The baby was started empirically on anti-failure treatment which consisted of furosemide and spironolactone.

Frontal chest radiograph showed cardiomegaly, while an echocardiography revealed mildly dilated left atrium and ventricle. A small persistent patent foramen ovale with left to right shunt was noted. There was also moderate tricuspid regurgitation. The ejection fraction was 68%. An abnormal aorto-pulmonary connection between ascending aorta and right pulmonary artery was demonstrated with left to right shunt.

CT angiography showed an abnormal tubular communication between the ascending aorta and the pulmonary trunk (**Figure 1**). This tubular communication was 6.6 mm in length with an entrance diameter of 5.4mm and exit diameter of 6.6 mm (**Figure 2**). She subsequently underwent surgical

repair at 2 months of age, which consisted of double ligations of the tubular connection through an open surgery.

Intraoperative findings showed an abnormal 'interconnecting vessel' between the aorta and main pulmonary trunk at the level of sinotubular junction which was then ligated. The aorta was found to be smaller than the main pulmonary trunk. A large patent ductus arteriosus (PDA) was ligated as well. The heart was moderately enlarged with moderate amount of pericardial effusion. The coronary vessels were unremarkable. The child was discharged well after 7 days in intensive care. Apart from several episodes of surgical wound infections which required antibiotics therapy, no major post-surgical complications were encountered.

Upon further follow-up at approximately two and a half years old, she was thriving well with no respiratory symptom. On auscultation, the lungs were clear with normal heart sounds and absent of any heart murmur. The ejection fraction remained stable at 67% on follow-up echocardiogram, without evidence of residual APW or PDA. There was no evidence of shunting, pulmonary arterial hypertension, or pericardial effusion.

DISCUSSION

Aortopulmonary window (APW) results from fusion failure of the conotruncal ridges (aorticopulmonary trunk) during the 5th to 8th week of gestation forming an abnormal communication between the proximal aorta and the main pulmonary artery (1). It usually begins just above the sinus of Valsalva with variable extensions to the aortic arch (2).

More than half of the APW cases may be found in isolation, while the remaining are found to be associated with other cardiac abnormalities. The most common associated cardiac anomalies are arch abnormalities, atrial septal defect, Tetralogy of Fallot and aortic origin of the right pulmonary artery. Other rare associations include tricuspid atresia, pulmonary or aortic atresia, ventricular septal defect and transposition of great arteries (2). In other literature, APW has been described as either simple or complex. Simple APW is associated with haemodynamically insignificant congenital cardiac anomalies, while Complex APW is associated with complex anomalies such as Tetralogy of Fallot, interrupted aortic arch, transposition of great arteries, or anomalous coronary arteries (3).

Patient with APW has unrestricted left-to-right shunt that worsens in post-natal period as the pulmonary vascular resistance falls dramatically. It results in congestive heart failure, pulmonary vascular obstructive disease and low cardiac output which may manifest as tachypnoea, diaphoresis, poor feeding, and inadequate weight gain. As the pulmonary vascular resistance increases rapidly, these patients are particularly susceptible to Eisenmenger's syndrome at an early age (3,4).

Mori's classification has been used in classifying APW. Type I APW occurs in the proximal part of aortopulmonary septum while type II defect occurs in the distal part of aortopulmonary septum, adjacent to the right pulmonary artery. Type III defect is a combination of both type I and II involving the entire length of the pulmonary trunk from immediately above the semilunar valves to the level of pulmonary bifurcation and the proximal portion of the right pulmonary artery (5).

Rather than having a typical window between the ascending aorta and pulmonary trunk, our case demonstrates a connecting tubular channel. Such tunnel-like configuration was reported by Ho et al. (6). The authors found a tunnel-like APW in 1 of the 25 patients who underwent transcatheter APW closure. The tunnel-shaped communication connected the right aortic sinus and the pulmonary sinus with the origin of the right coronary artery within the aortic origin (6).

Another resemblance to our findings was documented in a case reported by Chidambarathanu et al. (7). A 7mm tubular channel connected the proximal ascending aorta and the pulmonary trunk was described as tubular APW. Rather than a tubular APW, we propose the term aorto-pulmonary tunnel due to its simplicity and aptness.

An early and accurate diagnosis are of utmost importance. Imaging plays an important role in diagnosis as it is difficult to diagnose APW especially when it co-exists with VSD (3). APW should be suspected when there is a 'T-sign' in the presence of two normal semilunar valves, downward flow through the right side of main pulmonary artery and diastolic flow reversal in both aortic arch and descending aorta on echocardiography (4). Although 2-dimensional echocardiography is important in diagnosis, angiogram is considered the gold standard

for confirmation (3). Meanwhile, CT angiography provides precise anatomical details such as the location of the defect and may reveal other associated coronary origin anomalies (4). Furthermore, CT angiography is fast, safer, and less invasive compared to the transcatheter angiography.

Surgical closure is indicated in all patients diagnosed with APW except for asymptomatic patient with small APW. Surgical closure is usually performed at the time of diagnosis to reduce the risk of early pulmonary vascular disease (3). Due to the tubular nature of the APW communication, double ligations technique was able to be performed, rather than the conventional patches technique which required division of the defect prior to the patch repair.

CONCLUSION

Aortopulmonary window develops due to fusion failure of the conotruncal ridges. However, tubular structure or channel that connects proximal aorta with pulmonary trunk does not fit the typical APW. Early diagnosis and surgical closure are essential to prevent congestive heart failure and pulmonary vascular obstructive disease. CT angiography provides excellent visualization of the defect location and the origin of coronary arteries which enables appropriate surgical planning.

STATEMENT OF ETHICS

Written informed consent was obtained from the patients for publication of this case and any accompanying images.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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DATA AVAILABILITY STATEMENT

No additional data than the one presented in this article was used

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FIGURE LEGENDS

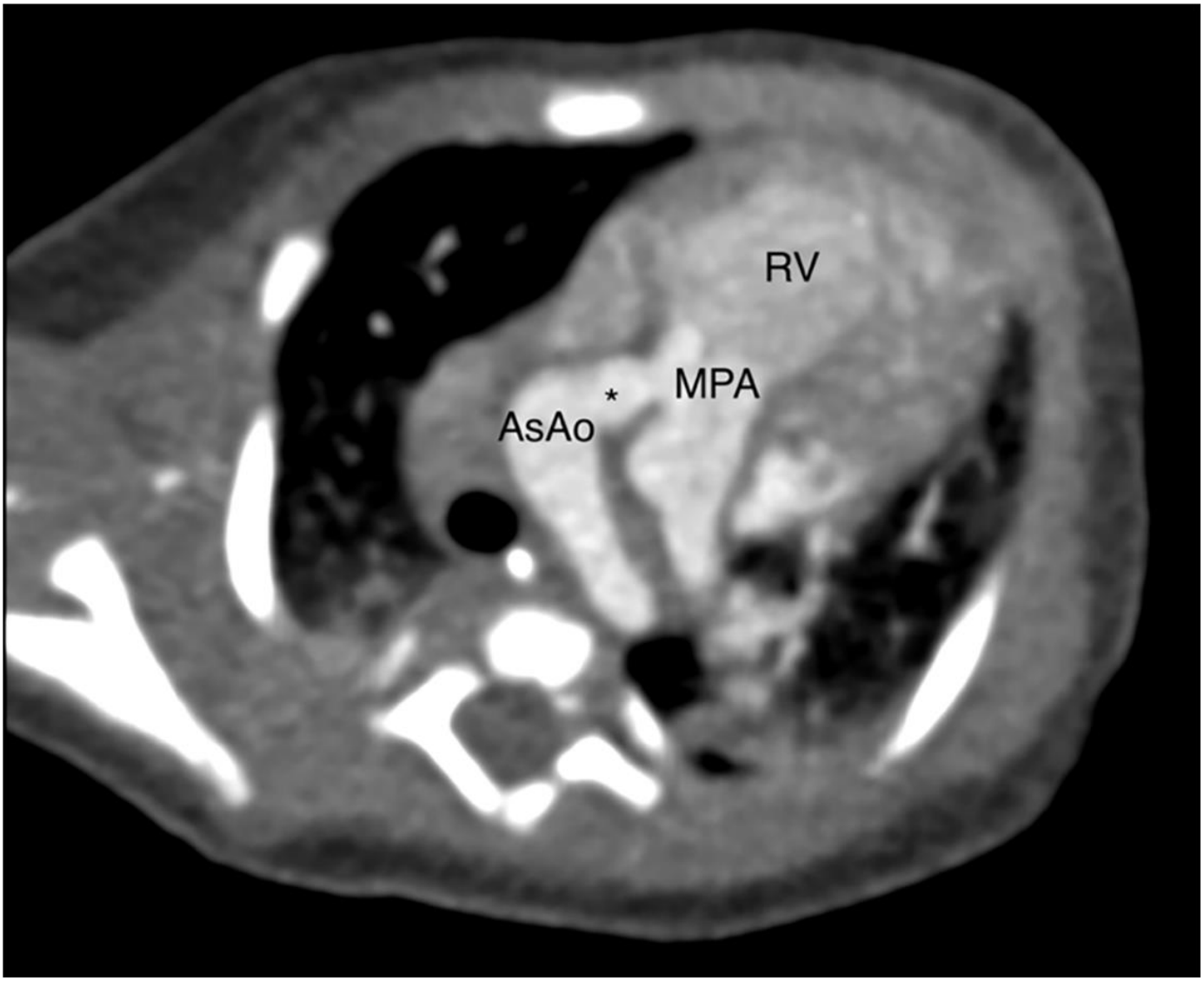

Figure 1: CT Pulmonary Angiogram in axial view showing an aortopulmonary window (*) that connects the main pulmonary artery (MPA) and ascending aorta (AsAo). Note that this communication appears as a tubular structure. RV: right ventricle.

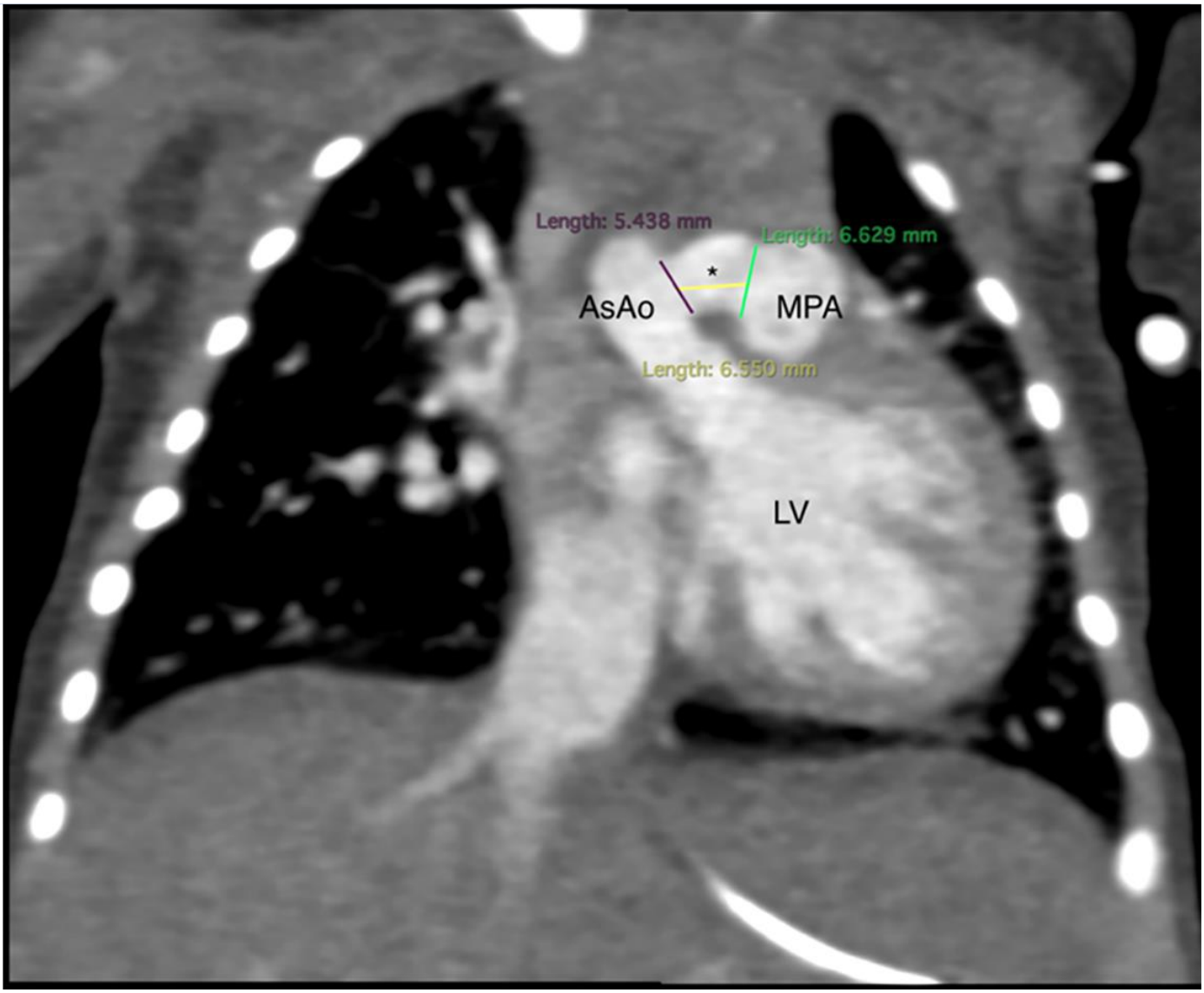

Figure 2: CT Pulmonary Angiogram in coronal view demonstrating an aortopulmonary window (*) that connects the main pulmonary artery (MPA) and ascending aorta (AsAo). Note the length of this tubular channel measures 6.6mm in length (yellow line). Its entry point (purple line) measures 5.4mm in diameter while the exit point (green line) measures 6.6mm in diameter. LV: left ventricle.