

Rare Case of Obstructed Supra Cardiac Total Anomalous Pulmonary Venous Connection to Hemiazygos Vein

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Abstract

This is a rare case of a neonate with obstructive supra cardiac total anomalous pulmonary venous connection at the level of the hemiazygos vein crossing the midline to join azygos vein to superior vena cava, where it was sandwiched between the spine and thoracic aorta. The diagnosis was elucidated with the advanced cardiac image (cardiac CT). The connection and the obstruction are rare and unique and could not be figured out easily with echocardiography.

Keywords: Pulmonary Venous; Supra Cardiac; Obstruction; Hemiazygos; Congenital Heart

Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease that accounts for 1% to 5% of cases [1]. 1953 Edwards JE the first to report the TAPVC to the azygos vein and later the prevalence of these subtypes in single institute case series around 6% [2,3].

The connection to the azygos vein was stenosis in all the reported cases in literatures [4,5].

Our case is rare and the connection to hemiazygos then crossing the midline to azygos vein before connecting to right superior vena cava. It was suspected as infracardiac total anomalous from the clinical presentation and initial echocardiography finding that later clarified with cardiac computed tomography (cardiac CT).

Case Presentation

- 37 weeks of gestation delivered by normal vaginal delivery as the second to a normal previous child born to non-consanguineous parents.
- Presented in the fourth day of life with Jaundice, poor intake and mild Tachypnea, soft systolic murmur, no clear cyanosis, the oxygen pulse oximeter had low oxygen saturation of 80-85% in room air.
- chest x-ray of the case showed marked pulmonary congestion and cardiomegaly.
- Echocardiography was done few times and final echocardiogram revealed that pulmonary veins (PVs) drain to confluence vein Crossing behind the midline to drain into right superior vena cava (SVC) supracardiac TAPVC, dilated SVC, dilated right atrium

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and ventricle, stretched patent foramen ovale and right to left shunt, small left atrium and compressed left ventricle.

- Cardiac computed tomography (CT) revealed Supracardiac-type TAPVC, the confluence of pulmonary venous drain connected to hemiazygos and crossing the midline between aorta and vertebra from left to right to azygos vein and finally to right superior vena cava with obstruction of pulmonary venous return figure 1 A-G.
- Surgical repair for TAPVC was performed urgently after establishing the diagnosis. The post-Operative course was smooth with progressive improvement of lung congestion.

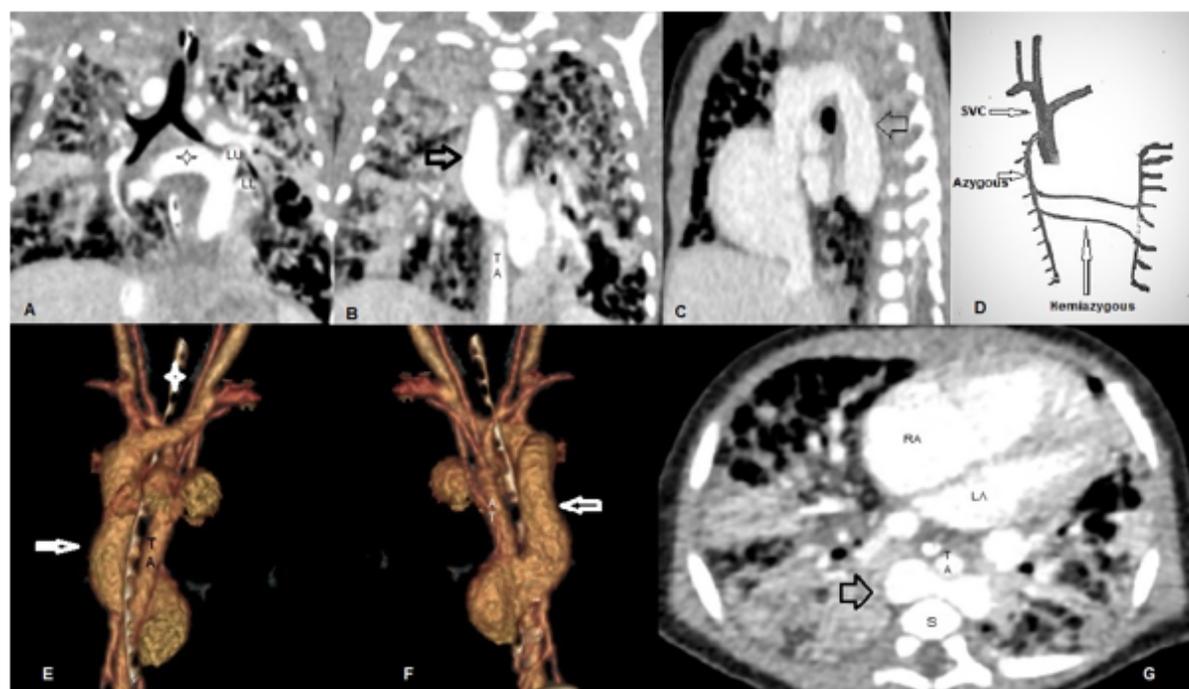


Figure 1: (A) cardiac CT angiogram: coronal view at the level of pulmonary veins showing the confluent, LL = Left Lower Pulmonary Veins, LU= Left Upper Pulmonary Vein, the right pulmonary veins confluent(star) and join the left pulmonary veins. (B) another coronal view of the chest showing the hemiazygos vein crossing behind thoracic aorta (TA) and anterior to spine (C) Sagittal view: azygos vein dilated and connected to superior vena cava (arrow) (D) Diagram showing hemiazygos and azygos and crossing of midline (E, F) Three-dimensional volume rendering cardiac CT: showing dilated azygos vein (arrow), compressed hemiazygos vein, Nasogastric tube (Star) (G) Axial view of the Cardiac CT showing the sandwiching of hemiazygos vein between spin (S) and thoracic aorta (TA), also the right heart, right atrium (RA) and right ventricle dilated and the left atrium (LA) small and underfill, most the image showed lung atelectasis and edema.

Discussion

- Various classifications of TAPVC were proposed but the most currently used by Darling et al according to the anatomic level of connection to draining systemic veins, supra, infra, and cardiac drain [8].

- In comparison to infra cardiac, supra cardiac TAPVC commonly has no obstruction, and if any obstruction usually where vertical vein compressed at the crossing between a left main bronchus and left pulmonary artery [6,7].
- Multiple levels of obstruction of the vertical vein in azygos connection were mentioned and our case has a unique drainage and obstruction site where pulmonary venous confluent connected to hemiazygos in the left side of midline and crossing to right where it trapped between aorta and spine adding another site for obstruction and drainage of supra cardiac TAPVC.
- The pulmonary venous Obstructive phenomena made the surgical interventional in this setting urgently and care higher than usual risk [7].

Conclusion

Total anomalous to hemiazygous vein is rare condition with has obstruction when it cross the midline from the left side of vertebral column to right side which add another mechanism of obstruction for supra cardiac type.

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