Journal of the Society for Cardiovascular Angiography & Interventions 3 (2024) 101303

JSCAI

The official journal of the Society for Cardiovascular Angiography & Interventions

JSCAI Image

Pulmonary Arteriovenous Malformation in an Unrepaired Tetralogy of Fallot: Diagnostic Implications

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Development of pulmonary arteriovenous malformation (PAVM) is not uncommon following Glenn repair for patients with congenital heart diseases; however, its occurrence with unrepaired tetralogy of Fallot (TOF) is rarely reported. The present case highlights this rare occurrence of PAVM in unrepaired TOF along with its potential diagnostic implications.

Computed tomography angiography images from an adult patient with TOF showed its classic imaging features, including the presence of subaortic ventricular septal defect with aortic override, infundibular and valvular pulmonary stenosis, and right ventricular hypertrophy. Pulmonary arteries were confluent with hilar right and left pulmonary arteries measuring 18 mm and 19 mm, respectively. Multiple significant aortopulmonary collaterals were seen arising from the proximal descending thoracic aorta, bilateral subclavian, and bilateral internal mammary arteries. Interestingly, it showed the presence of multiple collaterals along the right upper posterolateral chest wall (with hypertrophied and tortuous upper right posterior intercostal and lateral thoracic arteries) with the presence of PAVM in the superior segment (segmental artery diameter: 2.8 mm) of the right lower lobe (Figure 1). The aortic arch was right-sided with a mirror image branching pattern. Systemic and pulmonary venous drainage was normal. No patent arterial duct, anomalous coronary artery, or coarctation of the aorta was seen.

Pulmonary arteriovenous malformation is characterized by the presence of abnormal communication of pulmonary arteries and veins with no intervening capillary bed.[1] Most commonly, they are congenital in origin; however, they have been described in hepatic cirrhosis, post Glenn shunt, and rarely in mitral stenosis, constrictive pericarditis, and chronic thromboembolic pulmonary hypertension.[1–5] Although PAVM has been well described in cases of postoperative congenital heart diseases, including TOF (nearly one-fourth of cases following Glenn shunt), its occurrence in unrepaired TOF is rare.[6]

Its preoperative identification assumes great importance because it can be a cause of persistent hypoxemia (due to extracardiac right-to-left shunt) following TOF repair.[7] Moreover, it can be a cause of hemoptysis in such patients, where hemoptysis is generally attributed to the presence of hypertrophied bronchial or nonbronchial systemic collaterals.[8] Patients with TOF are more susceptible to serious neurologic complications, including stroke and brain abscess, which remain as the most commonly reported complications (stroke 18% and brain abscess 9%) of PAVM.[9,10] Its preoperative identification may allow an optimal treatment strategy to be developed prior to definitive intracardiac repair of TOF. If the PAVM is significant, one would expect postoperative cyanosis, at which point occlusion of the PAVM might be considered rather than focusing on lung parenchymal issues.

In the present case, it is difficult to determine if the PAVM is associated with the unrepaired TOF or if it is simply a second coexisting congenital abnormality, but this rare combination highlights the need for a thorough preoperative work-up as well as a high index of suspicion.

Keywords: cardiac computed tomography angiography; pulmonary arteriovenous malformation; tetralogy of Fallot.

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https://doi.org/10.1016/j.jscai.2024.101303
Received 27 October 2023; Received in revised form 9 January 2024; Accepted 10 January 2024
Available online 26 March 2024

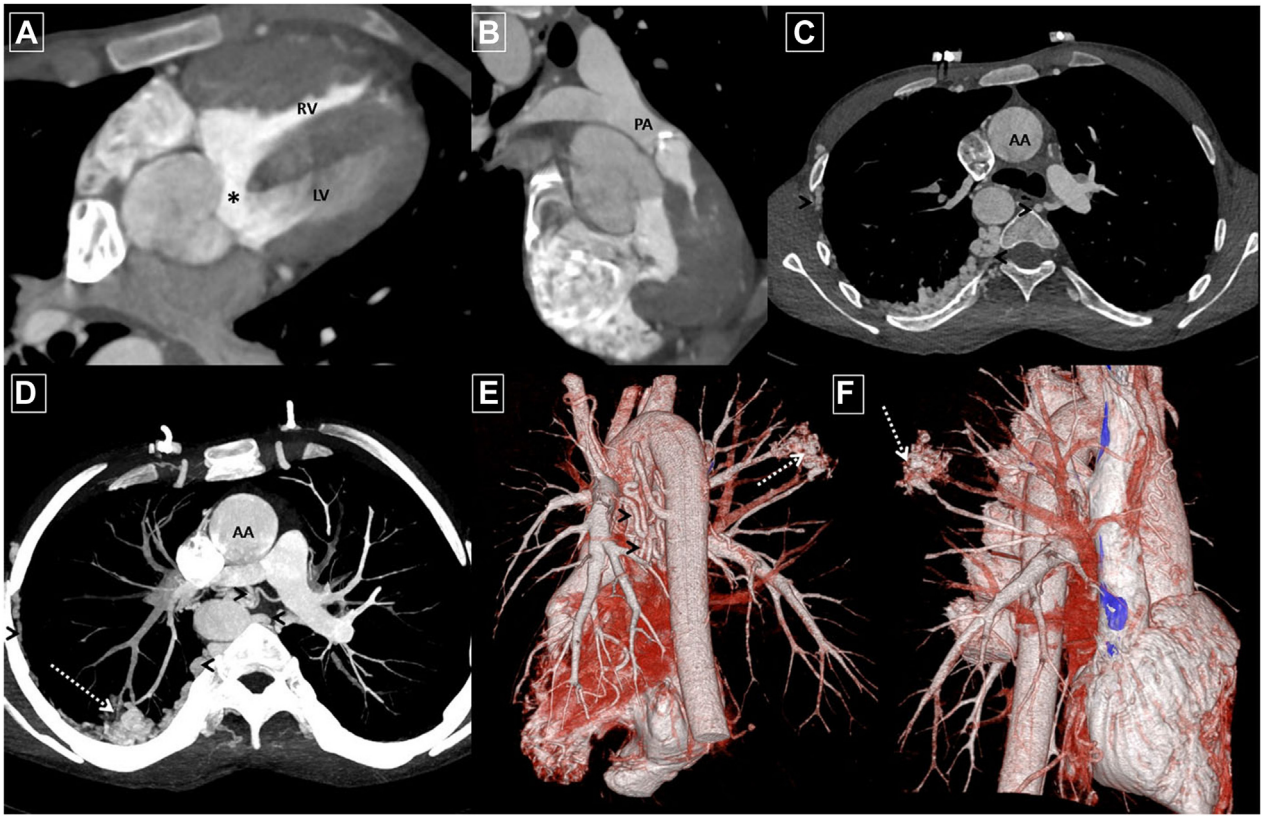

Figure 1.
Computed tomography (CT) angiography images from an adult patient with tetralogy of Fallot. (**A**) Subaortic ventricular septal defect (*) with aortic override and right ventricular hypertrophy (**B**) Infundibular and valvular pulmonary stenosis with valvular calcification and confluent pulmonary arteries. (**C, D**) Right-sided aortic arch with multiple major aorto-pulmonary collateral arteries (arrowheads), prominent bilateral internal mammary arteries, and the presence of pulmonary arteriovenous malformation (dashed arrow). (**E, F**) Volume-rendered CT images nicely depicting the pulmonary arteriovenous malformation (dashed arrow) with a demonstration of segmental feeding artery and the draining vein (in different densities) and the presence of significant aortopulmonary collaterals (arrowheads). AA, ascending aorta; LV, left ventricle; PA, main pulmonary artery; RV, right ventricle.

Declaration of competing interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding sources

This work was not supported by funding agencies in the public, commercial, or not-for-profit sectors.

Ethics statement and patient consent

Ethics approval was not applicable. Informed consent was obtained from the patient.

References

1. Meek ME, Meek JC, Beheshti MV. Management of pulmonary arteriovenous malformations. *Semin Intervent Radiol*. 2011;28(1):24–31. https://doi.org/10.1055/s-0031-1273937
2. Gossage JR, Kanj G. Pulmonary arteriovenous malformations. A state of the art review. *Am J Respir Crit Care Med*. 1998;158(2):643–661. https://doi.org/10.1164/ajrccm.158.2.9711041
3. Inami T, Yokoyama S, Seino Y, Mizuno K. Unique case of acquired pulmonary arteriovenous malformation developed by calcific constrictive pericarditis. *BMJ Case Rep*. 2013;2013. https://doi.org/10.1136/bcr-2012-008345
4. Chow LT, Chow WH, Ma KF. Pulmonary arteriovenous malformation. Progressive enlargement with replacement of the entire right middle lobe in a patient with concomitant mitral stenosis. *Med J Aust*. 1993;158(9):632–634. https://doi.org/10.5694/j.1326-5377.1993.tb137637.x
5. Sharma A, Gulati GS, Parakh N, Aggarwal A. Pulmonary arteriovenous malformation in chronic thromboembolic pulmonary hypertension. *Indian J Radiol Imaging*. 2016;26(2):195–197. https://doi.org/10.4103/0971-3026.184415
6. Bansal R, Talwar S, Ramakrishnan S. Simultaneous cardiac and pulmonary central cyanosis in a young girl: the double trouble. *Cardiol Young*. 2021;31(3):499–500. https://doi.org/10.1017/S1047951120004187
7. Kim SI, Park KY, Park CH, et al. Tetralogy of Fallot with pulmonary arterio-venous fistula. *Korean J Thorac Cardiovasc Surg*. 2000;33(3):257–261.
8. Haroutunian LM, Neill CA. Pulmonary complications of congenital heart disease: hemoptysis. *Am Heart J*. 1972;84(4):540–559. https://doi.org/10.1016/0002-8703(72)90479-6
9. White RI Jr, Lynch-Nyhan A, Terry P, et al. Pulmonary arteriovenous malformations: techniques and long-term outcome of embolotherapy. *Radiology*. 1988;169(3):663–669. https://doi.org/10.1148/radiology.169.3.3186989
10. Khurshid I, Downie GH. Pulmonary arteriovenous malformation. *Postgrad Med J*. 2002;78(918):191–197. https://doi.org/10.1136/pmj.78.918.191