

The first reported event of pulmonary venous communication to the esophageal mucosa

Authors: Bhoomi Dave, MD, Isabel Lopera, MD, Philip Araoz, MD

The Mayo Clinic, Rochester, MN

Purpose / outline

This abstract describes the first reported event of pulmonary venous communication to the distal esophageal mucosa in a patient

This unique vascular anomaly, identified through advanced cross-sectional imaging, resulted in pulmonary venous to systemic venous shunting without evidence of right ventricular volume overload or dysfunction.

The intersection of chronic esophageal inflammation and an anomalous pulmonary venous connection presents diagnostic and therapeutic challenges, as there is limited precedent to guide risk stratification or intervention.

Management required a multidisciplinary approach with the involvement of cardiology, interventional radiology, and gastroenterology expertise. Careful longitudinal surveillance was prioritized given the uncertain natural history and potential for hemodynamic or bleeding complications.

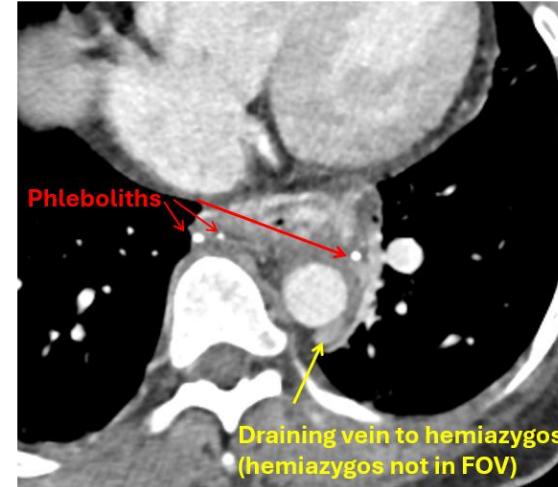
Because there are no established guidelines for managing this type of anomaly, the case highlights the need for individualized care, careful surveillance, and cross-specialty collaboration to address potential hemodynamic and bleeding risks.

Case presentation:

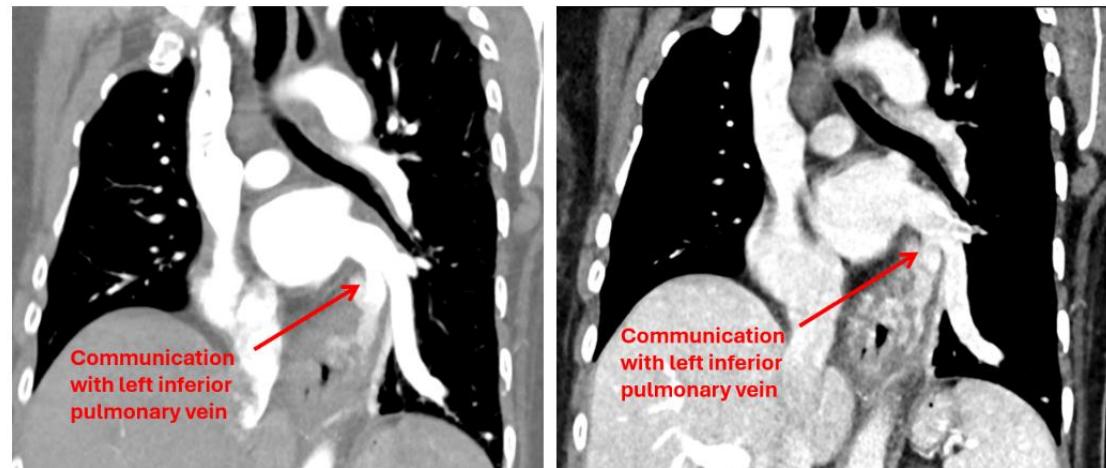
- A 45-year-old woman with recurrent esophagitis and a left lung base AVM presented with persistent, pressure-like chest discomfort
- Imaging revealed distal esophageal thickening, hiatal hernia, and a venous malformation of the distal esophagus supplied by the left inferior pulmonary vein, resulting in pulmonary venous to systemic venous shunting.
- Laboratory evaluation showed normal troponins, mild leukocytosis, and mild hypokalemia

A graded CT cardiopulmonary vein (CTCPV) demonstrated a venous malformation of the distal esophagus supplied by the left inferior pulmonary vein, therefore resulting in pulmonary venous to systemic venous shunting

Axial oblique



Coronal oblique



Arterial phase

20 seconds later (early venous phase)

Case presentation (continued):

- Laboratory evaluation showed normal troponins, mild leukocytosis, and mild hypokalemia.
- Clinically, the patient felt resolution of her chest pressure.
- Upon thorough review by the cardiology team, they indicated that the esophageal-venous malformation appeared to be low risk for acute complications.
- It was recommended she get serial echocardiograms and get close outpatient cardiology monitoring.

Discussion

- A direct communication between the cardiopulmonary vasculature and the esophagus via a venous malformation is extremely rare, with only isolated cases reported in the literature
 - Most previously described connections involve complex congenital anomalies or acquired fistulas, making isolated venous malformations of the esophagus a unique diagnostic challenge.
- Accurate diagnosis relies on advanced cross-sectional imaging modalities such as CT and MRI, which are essential for delineating vascular anatomy, guiding management, and differentiating these lesions from other entities

Discussion

- Clinical presentation varies from asymptomatic lesions to those causing pain, swelling, disfigurement, or functional impairment, with more than 60% of venous malformations progressing during adolescence, supporting consideration of early intervention.
- Diagnosis relies on clinical evaluation and imaging—ultrasound, MRI, and occasionally phlebography—to delineate lesion extent and differentiate from other vascular anomalies.

Management is best coordinated by a **multidisciplinary team**, with treatment tailored to lesion size, location, and symptoms.

Asymptomatic lesions may be monitored, while symptomatic or high-risk malformations are typically managed with **image-guided sclerotherapy or surgical excision**.



Discussion (continued)

- Risk stratification for gastrointestinal bleeding in esophageal venous malformations is informed by pressure gradient measurements.
- In cirrhosis, esophageal varices develop when hepatic venous pressure gradients exceed 10 mmHg, with bleeding risk increasing above 12 mmHg.
- While this patient does not have cirrhosis, consideration may be given to averaging proposed pressure gradients from echocardiogram to risk stratify the malformation with respect to bleeding risk.

Conclusion

- Consideration may be given to averaging proposed pressure gradients from echocardiogram to risk stratify the malformation with respect to bleeding risk. Intermittent echocardiogram could provide a noninvasive screening mechanism and help direct whether endoscopic visualization should be pursued
- This case demonstrates the role of radiology in identifying and characterizing rare vascular anomalies, directly influencing patient care by enabling precise risk stratification, and informing multidisciplinary treatment strategies

References

- 1. Cano Busnelli V, Medici J, Duro A, et al. Venous Malformation of the Esophagus. *Ann Thorac Surg.* 2018 Aug;106(2):e69-e71. PMID: 29626460. doi:10.1016/j.athoracsur.2018.03.006.
- 2. Yeh H, Wolf BS. A Pulmonary Venous Indentation on the Esophagus-a Normal Variant. *Radiology.* 1975 Aug;116(2):299-303. PMID: 1153729. doi:10.1148/116.2.299.
- 3. Reddy SM, Lander AD, Stumper O, et al. Esophago-Vascular Fistulae in Children: Five Survivors, Literature Review, and Proposal for Management. *J Pediatr Surg.* 2023 Oct;58(10):1969-1975. PMID: 37208288. doi:10.1016/j.jpedsurg.2023.04.014.
- 4. Legiehn GM, Heran MK. Venous Malformations: Classification, Development, Diagnosis, and Interventional Radiologic Management. *Radiol Clin North Am.* 2008 May;46(3):545-97, vi. PMID: 18707962. doi:10.1016/j.rcl.2008.02.008.
- 5. Dasgupta R, Patel M. Venous Malformations. *Semin Pediatr Surg.* 2014 Aug;23(4):198-202. PMID: 25241098. doi:10.1053/j.sempedsurg.2014.06.019.
- 6. La Mura V, Nicolini A, Tosetti G, Primignani M. Cirrhosis and portal hypertension: The importance of risk stratification, the role of hepatic venous pressure gradient measurement. *World J Hepatol.* 2015 Apr 8;7(4):688-95. doi: 10.4254/wjh.v7.i4.688.
- 7. Dubois J, Soulez G, Oliva VL, et al. Soft-Tissue Venous Malformations in Adult Patients: Imaging and Therapeutic Issues. *Radiographics.* 2001 Nov-Dec;21(6):1519-31. PMID: 11706222. doi:10.1148/radiographics.21.6.g01nv031519.