JACC: CASE REPORTS VOL. 29, 2024

© 2024 THE AUTHORS. PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN
COLLEGE OF CARDIOLOGY FOUNDATION. THIS IS AN OPEN ACCESS ARTICLE UNDER
THE CC BY-NC-ND LICENSE (http://creativecommons.org/licenses/by-nc-nd/4.0/).

IMAGING

IMAGING VIGNETTE: CLINICAL VIGNETTE

Coexisting Pulmonary Artery Sling and Persistent Left Superior Vena Cava



Amanda Naaraayan, MD, a Vitaly Buza, MD, PhD, a Christine Gerula, MD, a Alfonso H. Waller, MD, b

ABSTRACT

Congenital vascular anomalies such as pulmonary artery sling and persistent left superior vena cava are rare vascular disorders. We describe a case of a patient who presented with pneumonia and was found to have a pulmonary artery sling as a potential cause of the pneumonia. (J Am Coll Cardiol Case Rep 2024;29:102315) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

51-year-old man with past medical history of hypertension and class II obesity presented with shortness of breath, fever, chills, and a productive cough. A chest radiograph showed a right upper lobe infiltrate (Figure 1A) and he was diagnosed with pneumonia. The patient reported that this was his third diagnosis of pneumonia within the preceding 3 months. On further questioning, he also reported dysphagia. A computed tomography scan of the chest with contrast was performed and showed a left pulmonary artery arising from the right pulmonary artery (Figures 1B to 1D, Video 1) passing posteriorly to the trachea and anterior to the esophagus forming a pulmonary artery sling (PAS). The PAS was associated with some external compression of the posterior wall of the trachea above the level of the carina and external compression of the esophagus. Multiple patchy ground glass opacities were observed throughout both lung fields, which was interpreted as a multifocal pneumonia. The main pulmonary artery was dilated to 4.7 cm. In addition, there was a finding of a persistent left superior vena cava (PLSVC) which drained into a dilated coronary sinus (Figures 1B and 1E to 1F, Video 2).

PAS is a rare (0.14% of the general population) congenital anomaly created by aberrant origin of the left pulmonary artery from the posterior aspect of the right pulmonary artery. Presentation as a newborn or infant with severe respiratory symptoms is more typical and few are asymptomatic into adulthood. Literature review finds <30 adults newly diagnosed with PAS. It commonly has compressive effects on the trachea which could lead to respiratory symptoms such as obstructive pneumonia, atelectasis, emphysema, and stridor. External compression on the esophagus could lead to dysphagia or aspiration pneumonia. A PLSVC is also rare (0.3% to 3% of the general population) and typically discovered incidentally. PLSVC drains the left jugular and left subclavian veins. In 80% to 90% of cases, it drains into the right atrium via the coronary sinus, as observed in our patient. There are limited data regarding PAS and PLSVC being found concomitantly, and the

From the ^aDivision of Cardiology, Department of Internal Medicine, Rutgers New Jersey Medical School, Newark, New Jersey, USA; and the ^bDepartment of Radiology, Rutgers New Jersey Medical School, Newark, New Jersey, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received October 31, 2023; revised manuscript received February 26, 2024, accepted March 6, 2024.

ABBREVIATIONS AND ACRONYMS

PAS = pulmonary artery sling
PLSVC = persistent left
superior vena cava

most-described anomalies found alongside a PLSVC do not include PAS. Surgical repair is indicated for all symptomatic PAS.

Our patient had a history of recurrent pneumonia and was referred for bronchoscopy, an upper gastrointestinal series, and surgical evaluation for possible repair of the PAS.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Alfonso H. Waller, Rutgers New Jersey Medical School, 185 S Orange Avenue, Newark, New Jersey 07103, USA. E-mail: ahw22@njms.rutgers.edu. @alfonsowallermd.

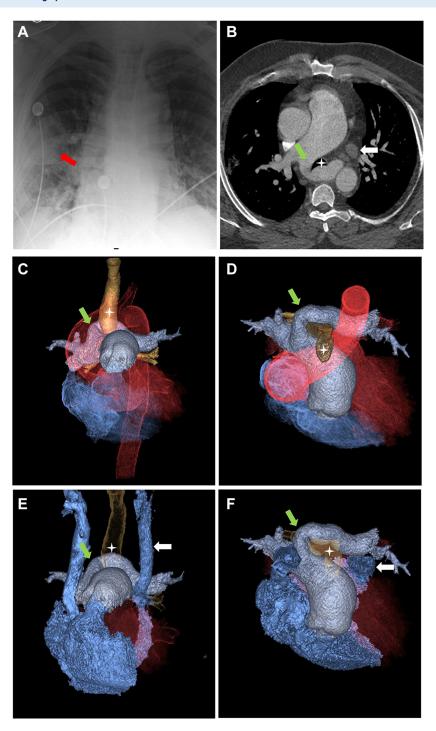
REFERENCES

- **1.** Xie J, Juan YH, Wang Q, et al. Evaluation of left pulmonary artery sling, associated cardiovascular anomalies, and surgical outcomes using cardiovascular computed tomography angiography. *Sci Rep.* 2017;7:40042.
- **2.** Sade RM, Rosenthal A, Fellows K, Castaneda AR. Pulmonary artery sling. *J Thorac Cardiovasc Surg.* 1975;69(3):333–346.
- **3.** LaBelle MF, Rainer WG, Ratzer E, Miller KB. Surgical repair of pulmonary artery sling in an adult. *Annals Thorac Surg.* 2010;90(3):1009-1011
- **4.** Tyrak KW, Holda J, Holda MK, Koziej M, Piatek K, Klimek-Piotrowska W. Persistent left superior vena cava. *Cardiovasc J Afr.* 2017;28(3): e1–e4.

KEY WORDS persistent left superior vena cava, pulmonary artery sling

APPENDIX For supplemental videos, please see the online version of this paper.

FIGURE 1 Chest Radiograph and Contrast-Enhanced CT



(A) Chest radiograph shows a right upper lobe consolidation (red arrow). (B) An oblique maximum-intensity projection computed tomography image shows the trachea (star), and the left pulmonary artery arising from the right pulmonary artery (green arrow) with a retro-tracheal course. Incidentally, a persistent left superior vena cava is seen (white arrow). (C) A volume-rendered image shows the trachea (star) and the left pulmonary artery arising from the right pulmonary artery (blue, green arrow) with a retro-tracheal course. (D) A volume-rendered image from a superior perspective shows the trachea (star) and the left pulmonary artery arising from the right pulmonary artery (blue, green arrow) with a retro-tracheal course. (E) A volume-rendered image shows the trachea (star), the left pulmonary artery arising from the right pulmonary artery (blue, green arrow) with a retro-tracheal course, and a persistent left superior vena cava is seen draining into the coronary sinus (white arrow). (F) A volume-rendered image from a superior perspective, which shows the trachea (star), the left pulmonary artery arising from the right pulmonary artery (blue, green arrow) with a retro-tracheal course, and a persistent left superior vena cava is seen draining into the coronary sinus (white arrow).