Aorta–to–Pulmonary Vein Fistula in an Asymptomatic 25-Year-Old Man

Arun Dahiya, MD; Patrick Collier, MB, PhD; Richard Krasuski, MD; Vidyasagar Kalahasti, MD; Pedro del Nido, MD; William J. Stewart, MD

A 25-year-old man with no significant medical history was found to have a continuous murmur along the left sternal border on routine physical examination performed in preparation for running a marathon. Echocardiography showed a dilated left ventricle with normal function and continuous high-velocity flow in an aberrant vessel in close vicinity to the abdominal aorta (Movie I in the online-only Data Supplement). Chest x-ray (posterior-anterior and lateral projections, Figure 1) disclosed conglomeration of tubular densities in a scimitar pattern in the right lower lobe suggestive of anomalous pulmonary venous return of right lower lobe (scimitar syndrome; anomalous drainage of right lung pulmonary veins into the inferior vena cava). Cardiac magnetic resonance imaging (MRI) not only demonstrated normal drainage of all 4 pulmonary veins into the left atrium but also revealed an aberrant vessel (Movie II in the online-only Data Supplement) extending between the abdominal aorta and the right inferior pulmonary vein with an elevated systemic-to-pulmonary flow ratio of 1.8, consistent with a large left-to-left shunt. Cardiac computed tomography (CT) confirmed the presence of a large fistulous connection between the abdominal aorta and the right inferior pulmonary vein (Figure 2 and Movie III in the online-only Data Supplement) located in the right lower lobe (Figure 3A). Although there was no distinct lung parenchyma changes on CT and MRI, the presence of conglomerated fistula vessels in the right lower lung lobe limited the ability to exclude associated intralobar lung sequestration. Therefore, cardiac catheterization (Movie IV in the online-only Data Supplement) was subsequently performed before consideration of percutaneous closure of the fistula. However, this treatment option was abandoned because of concerns that perfusion of the right lower lung lobe was fistula dependent. The patient was then referred for open surgery to determine whether the fistula could be closed safely, which was done by temporarily occluding the fistula with a snare and observing the color of the segment of the lung after 5 minutes of

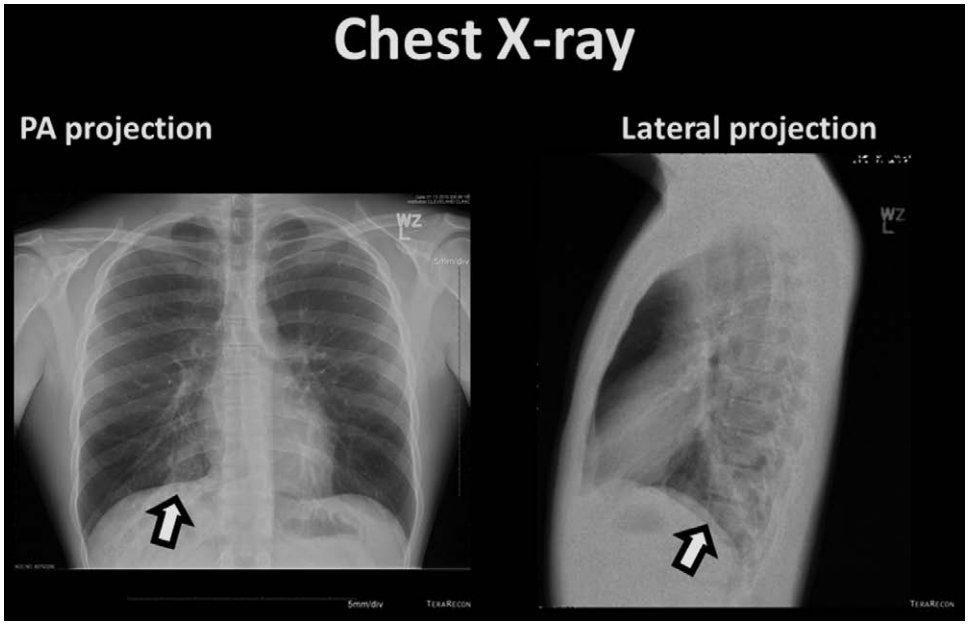

Figure 1. Chest x-ray (posterior-anterior and lateral projections) revealing conglomeration of tubular densities in a scimitar pattern in the right lower lobe (**arrow**).

From the Department of Cardiovascular Medicine, Cleveland Clinic Foundation, Cleveland, OH (A.D., P.C., R.K., V.K., W.J.S.); Department of Cardiovascular Surgery, Children Hospital Boston, MA (P.d.N.); School of Medicine, University of Queensland, Queensland, Australia (A.D.); and Royal Brisbane and Women's Hospital, Queensland, Australia (A.D.).

The online-only Data Supplement is available with this article at http://circ.ahajournals.org/lookup/suppl/doi:10.1161/CIRCULATIONAHA.112.125757/-/DC1.

Correspondence to Arun Dahiya, MD, Department of Cardiology, Royal Brisbane and Women's Hospital, Herston, Queensland 4029 Australia. E-mail arun_dahiya@health.qld.gov.au

(*Circulation*. 2013;127:1727-1729.)

***Circulation* is available at http://circ.ahajournals.org** **DOI: 10.1161/CIRCULATIONAHA.112.125757**

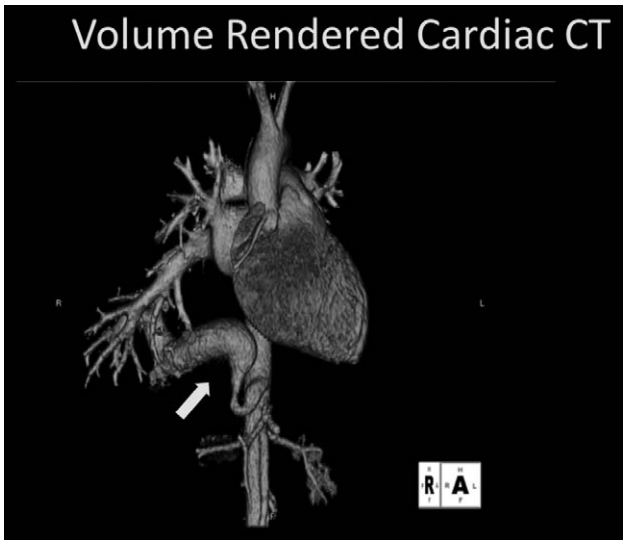

Figure 2. Volume-rendered cardiac computed tomography revealing the presence of a fistula (**arrow**) between the abdominal aorta and the right inferior pulmonary vein.

occlusion. Arterial blood gases were also obtained at that time. Because no changes were detected, the fistula was then permanently ligated. Although inspection of the lung surface during fistula occlusion is not a direct indication of the presence of normal pulmonary artery blood supply to that segment, it can confirm that pulmonary infarct is unlikely to occur. This was the only concern and the reason for the test occlusion. Ultimately, surgical ligation resulted in no complications, and there was no need for lung resection. Postoperative cardiac CT subsequently showed that the left ventricular size reverted to normal and that no residual fistula was present (Figure 3A and 3B).

An abdominal aorta–to–pulmonary vein fistula is a very rare congenital anomaly that previously had been described only in infancy.[1,2] To the best of our knowledge, this is the first case report of a large systemic circulation–to–pulmonary vein fistula associated with chamber dilation and a high-volume shunt in an asymptomatic adult. This case highlights the key role of multimodality imaging in the diagnosis and management of a rare congenital anomaly. Tomographic investigations such as cardiac CT or MRI should be considered in cases when echocardiography fails to identify the cause of a murmur, particularly if chamber dilation is present. More specifically, in the setting of a systemic–to–pulmonary vein fistula, cardiac CT or MRI can also diagnose the presence of concomitant lung sequestration,[3–5] which is critical with regard to subsequent disease management. Lung sequestration is defined as a nonfunctioning mass of normal lung tissue that lacks normal communication with the pulmonary arterial system and the tracheobronchial tree[3] and receives its blood supply from 1 or more anomalous systemic arteries. On the basis of its pleural covering, lung sequestration is further divided into 2 subtypes. Extralobar lung sequestration has a distinct pleural covering resulting in complete anatomic separation of the mass from adjacent normal lung tissue, and intralobar sequestrations are masses of lung tissue in continuation with adjoining normal lung, which was the

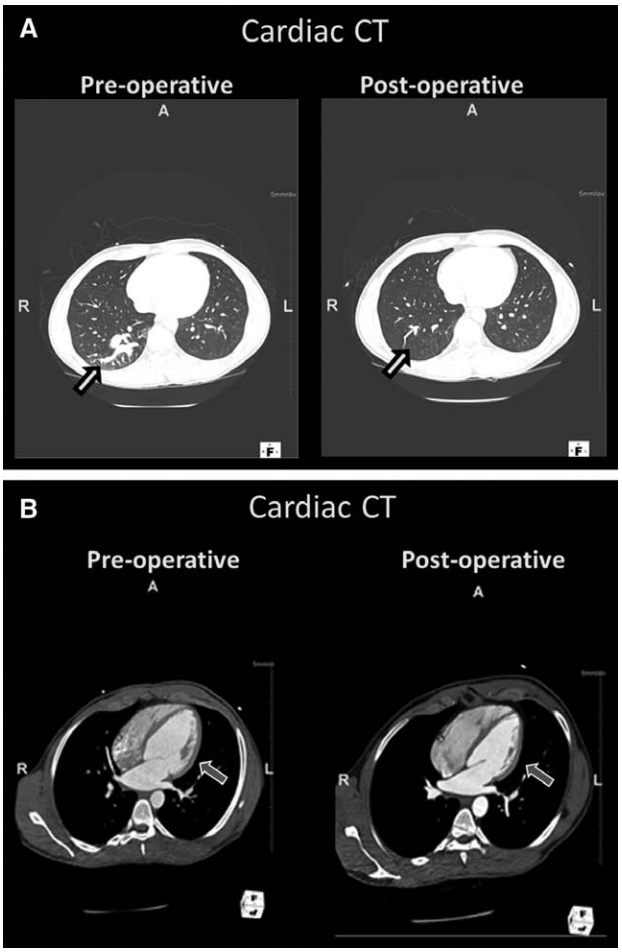

Figure 3. A, Presurgical cardiac computed tomography (CT) images demonstrating a fistula within the posterior right lower lung lobe (yellow **arrow**). Postsurgical cardiac CT shows normal lungs after successful ligation of the fistula. **B,** Presurgical cardiac CT images demonstrating dilated left ventricle (red **arrow**). Postsurgical cardiac CT shows a reduction in the size of the left ventricle after successful ligation of the fistula.

main consideration in our case. Lung sequestration if present usually requires lobectomy[6] or embolization.[7] In contrast, patients without sequestration can be managed by surgical ligation[1] or percutaneous closure of the fistula.[2] CT is excellent at depicting parenchymal abnormalities associated with lung sequestration, which has a typical appearance of a complex mass with and without cystic changes. CT angiogram and multiplanar MRI not only can demonstrate parenchymal lung changes but also can delineate its systemic arterial supply and venous drainage. Invasive angiography may be indicated if noninvasive evaluation is nonconfirmatory.[8] Rarely, in patients with hemodynamically significant aberrant pulmonary vessel, noninvasive and invasive evaluation is unable to confidently rule out associated lung sequestration (as in this case), warranting the safer approach of surgical ligation because it facilitates direct inspection of the lung surface for infarction during temporary fistula occlusion, which in turn dictates the final surgical strategy.

Disclosures

None.

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