A 14-month-old girl was evaluated for noisy breathing and swallowing. An upper gastrointestinal barium swallow study showed a filling defect of the esophagus at the level of the aortic arch suggestive of a vascular ring (Figures 1 and 2). A chest computed tomography angiogram with 3-dimensional reconstruction (1-mm sections; Movies I and II in the online-only Data Supplement) showed a vascular ring consisting of a left aortic arch, retropharyngeal transverse aorta, aberrant right subclavian artery, right descending aorta, and a small right-sided patent ductus arteriosus to the right pulmonary artery (Figure 3). This is an extremely rare form of vascular ring because of the presence of the right-sided patent ductus arteriosus. A left aortic arch and aberrant right subclavian artery are often considered a normal variant and not a true vascular ring because of the typical absence of a right-sided ductus arteriosus or ligamentum arteriosum. In this case, the ductus arteriosus was patent, allowing clear visualization on cross-sectional images. Often, however, this is not the case, and other indirect findings must be used for diagnosis of the presence of a ligamentum that would complete a vascular ring. As shown here, rightward tethering of the vascular structures (aberrant right subclavian and right ductus/ligamentum) may result in a “circumflex” course of the isthmus and proximal descending aorta. In addition, a Kommerell diverticulum (not seen in this case) denotes the presence of a ligamentum arteriosum. Delineation of vascular anatomy with high-resolution imaging was helpful for diagnosis and surgical planning. This patient underwent a successful surgical division of the vascular ring and reimplantation of the right aberrant subclavian artery via a right thoracotomy.

Disclosures

None.

References

Figure 1. Frontal still image of the upper gastrointestinal series after oral contrast was given. There is a large oblique filling defect at the level of the left aortic arch.

Figure 2. Lateral still image of the upper gastrointestinal series after oral contrast was given. There is a large posterior filling defect at the level of the left aortic arch.

Figure 3. Cross-sectional still image of the patient’s chest computed tomography angiogram with the arrows pointing to the anatomic structures at the level of the vascular ring such as the ascending aorta (Asc. Ao.), right pulmonary artery (RPA), left pulmonary artery (LPA), patent ductus arteriosus (PDA), and retropharyngeal transverse aortic arch.
Left Aortic Arch, Right Descending Aorta, and Right Patent Ductus Arteriosus: Precise Depiction of a Rare Vascular Ring With Cardiac Computed Tomography
Hoang H. Nguyen, Elizabeth Sheybani, Peter Manning, Tom Herman and Shafkat Anwar

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