Dysphagia is a common presenting condition in outpatient otolaryngologic care. Most physicians have a well-established algorithm for evaluating dysphagia that begins with a thorough history and physical examination. After initial evaluation, most otolaryngologists obtain functional imaging in the form of a barium swallow. Although a vascular cause, such as dysphagia lusoria, is uncommon, the use of standard imaging techniques makes it unlikely that vascular causes will be missed. Furthermore, physicians involved in the evaluation and treatment of dysphagia should not only be aware of this diagnosis but also understand the appropriate evaluation and treatment.

The first documented case of dysphagia lusoria was described in 1794 by Bayford,1 who termed the compression of the esophagus by an aberrant subclavian artery lusus naturae or freak of nature. An aberrant right subclavian artery occurs in 0.5% of the population, whereas an aberrant left subclavian artery is a more unique finding, occurring in 0.06% to 0.01% of the population.2 The overall rarity of dysphagia lusoria was quantified in a recent study, which suggested that only 0.36% of patients with dysphagia had aberrant subclavian arteries.3

Report of a Case
A woman in her 20s with no significant medical history presented with 2 months of progressive dysphagia to solid foods. She reported daily regurgitation 1 to 2 hours after meals. She denied any difficulty swallowing liquids or a history of similar problems. She denied any stridor, wheezing, coughing, or shortness of breath. She denied any systemic symptoms, including unintentional weight loss. Initial physical examination findings were unremarkable.

On the basis of the patient’s presenting symptoms, a barium swallow was obtained, which demonstrated a narrowing of the esophagus secondary to posterior extrinsic compression (Figure 1). Follow-up chest radiography revealed a right-sided aortic arch with soft tissue attenuation just superior to the left pulmonary artery (Figure 2). Further evaluation by computed tomography revealed an aberrant left subclavian artery with an associated Kommerell diverticulum (Figure 3). Preoperatively, the patient underwent angiography to further define the anatomy, confirming the prior anatomical findings and illustrating a vascular ring formed by a persistent ligamentum arteriosum (Figure 4).

The patient underwent a left subclavian to left carotid transposition via a supraclavicular incision, division of the diverticulum, and subsequent resection of the ligamentum arteriosum via left thoracotomy. At the patient’s 1-month follow-up appointment, she reported full resolution of her presenting symptoms.

Discussion
Aortic arch anomalies account for 1% to 3% of congenital heart disease.4 In cases where the arch crosses the right main
bronchus, it is termed a right aortic arch. When this occurs, the aorta most commonly descends along the right side of the spine. This anatomical variant alone will not lead to dysphagia. Compression of the esophagus may occur when both the ligamentum arteriosum and a Kommerell diverticulum are present. The ligamentum completes the vascular ring that connects the diverticulum posteriorly behind the esophagus to the left pulmonary artery anteriorly. Kommerell diverticulum, a remnant of the left dorsal arch, is located where the left aberrant subclavian artery originates, causing external posterior compression of the esophagus.
Patients often become symptomatic in later decades as the arteries become tortuous and aneurismal.4

The patient described in this case is unique not only because she had both a right aortic arch and aberrant left subclavian artery but also because she presented 3 decades earlier than expected. Her diagnosis most likely would have been delayed had a vascular cause not been suspected due to the findings on her barium swallow. As was visible on her study, shallow oblique posterior compression of the esophagus is a hallmark sign for an aberrant subclavian artery and should prompt the physician to obtain a chest radiogram.5 Once diagnosis of a vascular ring or aberrant subclavian artery is determined, referral to vascular surgery is warranted for further treatment.

The first documented surgical repair of subclavian artery aberrancy was performed by Robert Gross in 1946 and consisted of simple ligation of the aberrant vessel at its origin.6 A drawback of this surgery was left or right arm weakness dependent on the location of the aberrancy.7 In 1965, the surgical approach was modified to reconstitute blood flow to the subclavian artery via transposition of its distal aspect to the carotid and is now considered standard of care.8 In young patients, division of Kommerell diverticulum is warranted to prevent additional surgery for recurrent dysphagia.9

Conclusions

Because dysphagia is a common presenting condition, it is important to have a well-defined algorithm for evaluating patients. Although uncommon, vascular causes should remain in the differential diagnosis. When extrinsic compression of the esophagus is noted on barium swallow, further evaluation and testing are advised to determine whether there is a vascular component to the compression. Surgical correction of the anomaly will typically lead to resolution of the patient’s symptoms.

REFERENCES