A 42-year-old woman who had a diagnosis of asthma since childhood that was refractory to corticosteroid therapy complained of chest discomfort, progressive dyspnea on exertion, and chronic cough, all gradually worsening in severity over the past several years. Her chest pain was described as a constant band-like pressure encircling the chest wall, and was exacerbated by cold weather, exertion, supine position, and forced expiration. The chest pain occasionally radiated to the throat and left shoulder. Both the chest pain and dyspnea were partially relieved by inhaled β-agonist, nitroglycerin, and/or rest. In addition, she complained of intermittent dysphagia for solids and liquids. Her medications included a short-acting β2-agonist, inhaled corticosteroids, and intermittent oral corticosteroids.

Results of her physical examination were within normal limits, except for mild Cushingoid features. Routine laboratory blood studies were unremarkable. A chest radiograph is shown in Figure 1. Findings on ECG, cardiac enzymes during a severe episode of chest pain, two-dimensional echocardiogram, cardiac catheterization, and a 24-h ambulatory pH probe were all within normal limits. Spirometry showed a FVC of 3.67 L (102% of predicted), FEV1 of 2.96 L (99% of predicted), and FEV1/FVC ratio of 81%. The flow-volume loop showed flattening of the expiratory segment (Fig 2).

What is the cause of this patient’s chest pain, dyspnea, and dysphagia?
Figure 1. The chest radiographs of a 42-year-old women with asthma-like symptoms revealing a right-sided aortic arch and distal trachea narrowing.

Figure 2. Flow-volume loop demonstrating flattening of the expiratory portion, suggesting variable intrathoracic obstruction. Pred = predicted.
Diagnosis: Vascular ring leading to symptomatic tracheoesophageal compression

The chest radiograph showed a right-sided aortic arch and distal tracheal indentation (Fig 1). A thoracic CT scan also revealed a right-sided aortic arch with a right descending thoracic aorta, a fibrous band, and an aberrant left subclavian artery (Fig 3). In addition, a retroesophageal diverticulum produced anterior displacement and compression of the trachea and esophagus. Bronchoscopy revealed tracheal compression 3 cm above the carina and extending proximally 3 cm in length. Because of persistent symptoms from the vascular ring, a left posterolateral thoracotomy through the fourth intercostal space was performed. At surgery, the ligamentum arteriosum was identified as a thick, fibrous band compressing both the esophagus and trachea, and an aberrant left subclavian artery was also seen traveling behind the esophagus. The band was divided. Postoperatively, the patient had immediate resolution of her chest pain and dyspnea. Her dysphagia also improved. She has remained asymptomatic since the operation, and has not required corticosteroids or any other asthma medications.

A vascular ring is a rare congenital abnormality that can cause complete or partial obstruction of the esophagus and/or trachea. A complete vascular ring is formed by a right aortic arch, left ligamentum arteriosum that fails to regress, and aberrant left subclavian artery. Vascular rings occur with an incidence of 0.2%, and usually presents in infancy or childhood with respiratory symptoms consisting of wheeze, recurrent respiratory tract infection, or stridor. The mean age at onset of symptoms ranges from 7 months to 4.5 years. GI symptoms such as emesis and/or dysphagia occurred in approximately 40% of cases. Most patients with vascular rings will require surgical repair. Tracheoesophageal compression has been described in a large series of infants and children, but documented cases in adults are rare. While the incidence of vascular rings in adults is not known, the occurrence of right-sided aortic arch, an anomaly associated with a higher likelihood of a complete vascular ring, is approximately 0.1%. In adults, either swallowing or respiratory symptoms may predominate with the presence of a vascular ring.

The right aortic arch with ligamentum arteriosum form of vascular ring causes between 25% and 30% of all cases of vascular rings. In fact, it is the second most common type of vascular ring requiring surgery, occurring in an estimated 1 in 1,000 individuals in the general population. In this type of abnormality, as seen in our patient, the aortic arch is to the right of the trachea. The ligamentum arteriosum extends from the main pulmonary artery to the descending thoracic aorta, completing the ring. The right aortic arch with ligamentum arteriosum form of vascular ring results from a persistent right fourth brachial arch and an absent left arch, which is interrupted between the left common carotid and left subclavian artery. Isolated aortic arch abnormalities have been associated with chromosome 22q11 deletion, and 75% of patients with this chromosomal abnormality have congenital cardiovascular defects.

The respiratory symptoms associated with this anomaly may result from either static or dynamic changes in the trachea, bronchi, lungs, and thoracic cage. Vascular rings tend to become more constrictive as the trachea grows. The tracheomalacia may diminish once the compression is relieved. Symptoms may occur transiently with exertion, supine positioning, and fluid administration that dilates the aorta. Superimposed illnesses, such as asthma or respiratory tract infections, may cause further increases in airway resistance exacerbating airflow obstruction. In addition, gastroesophageal reflux due to the esophageal compression from the vascular ring may also contribute to the respiratory symptoms.

The diagnostic workup for this anomaly should include a chest radiograph, which will often show a right-sided aortic arch, a distal tracheal indentation on the posterolateral view, and a retrotracheal opacity or anterior tracheal bowing on lateral view. A barium swallow may also show compression of the esophagus. Additional studies that might be...
helpful include a thoracic CT scan or MRI, and an aortogram. Two-dimensional echocardiography has been reported to be insensitive in the diagnosis of vascular rings. The value of bronchoscopy is primarily to rule out other anatomic causes of airway obstruction, including endobronchial neoplasm or airway stenosis. Spirometry findings may be normal or may show flattening of the expiratory portion of the flow-volume loop, suggesting a variable intrathoracic obstruction. The diffusion capacity is typically normal.

Surgical division of the ligamentum arteriosum is the treatment of choice in symptomatic patients. In those who continue to have persistent symptoms despite surgery, tracheal resection or intraluminal stenting may be necessary.

Vascular rings should be included in the differential diagnosis of all adult patients with dyspnea and/or dysphagia accompanied by an aortic arch anomaly. Diagnosis of this condition in patients with chronic, nonspecific respiratory complaints may eliminate the need for corticosteroids and other respiratory medications. A high degree of clinical suspicion is required for the recognition of this rare disease entity.

References
7 Hastreiter AR, D’Cruz IA, Canatez T, et al. Right-sided aorta.