Aberrant right subclavian artery syndrome manifesting as focal tracheomalacia

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ABSTRACT

Tracheomalacia is one of the rarest etiologies of chronic cough. Herein, we present the case of an elderly woman with chronic cough who on detailed evaluation was found to have focal (localized) tracheomalacia due to compression by an aberrant right subclavian artery (arteria lusoria). The absence of dysphagia in this patient also deserves particular mention.

KEY WORDS: Aberrant right subclavian artery, focal, tracheomalacia

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THE CASE

A 59-year-old woman was referred to us for the evaluation of dry cough, which she had for the past 9 years. She described her cough as dry which occurred in paroxysms and was more at night. Her vital signs were normal, and examination of the chest was unremarkable. Routine blood investigations were within normal limits, and sputum studies for mycobacterial as well as bacterial infections were negative. Chest X-ray posteroanterior

view did not reveal any abnormality. Because of the brassy nature of her cough, a possible tracheal pathology, in particular, tracheomalacia was considered. A contrast-enhanced computed tomography (CECT) scan of the neck and chest was taken with both inspiratory and expiratory scans.

QUESTION

What is the diagnosis?

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ANSWER

Focal tracheomalacia due to compression by aberrant right subclavian artery. $^{[1]}$

DISCUSSION

The CT scan findings were consistent with a diagnosis of tracheomalacia. The axial inspiratory and expiratory films [Figure 1] demonstrated significant tracheal narrowing of >50% during expiration. Very interestingly, the CECT also showed an aberrant subclavian artery arising from the distal aortic arch, and in its retroesophageal course, compressing on the tracheal lumen at T2 vertebral level [Figures 2 and 3]. The sagittal diameter of the tracheal lumen just above the level of aberrant right subclavian artery measured 1.40 cm on inspiratory phase and 0.42 cm on expiratory phase, with a 70% reduction in diameter. Thus, the final diagnosis of aberrant right subclavian artery causing localized tracheomalacia was made. At this juncture, the patient was interrogated again in detail as to whether she had dysphagia at any point of time, but she denied having the same. The patient was offered vascular surgical correction, but she declined. She was continued on symptomatic management with cough suppressants and reported improvement in symptoms.

Tracheomalacia is defined as segmental or diffuse weakness of the trachea. [2] If the main stem bronchi are also involved, the term tracheobronchomalacia is used. It often results in exaggerated narrowing of the tracheal lumen during expiration and widening during inspiration. [3] Tracheomalacia can be classified in various ways. Depending on the shape of trachea, it could be cresent, lateral, or circumferential. Based on degree of involvement, it could be segmental or diffuse, tracheal, bronchial, or tracheobronchial. The most commonly accepted classification is as congenital or acquired. [4]

Acquired tracheomalacia is more common than congenital. Causes include tracheostomy, endotracheal intubation, tracheal cartilage injury, relapsing polychondritis, and chronic tracheal compression from goiter, tumors, cysts, or vessels.^[4,5] The congenital variety may occur in isolation or may be found along with other defects such as laryngomalacia and laryngeal clefts.^[6] Because they are unable to expel out secretions, patients with tracheomalacia present with chronic cough, dyspnea, recurrent respiratory infections, and bronchiectasis.^[7]

Aberrant right subclavian artery (also called arteria lusoria)^[1] is assumed to be the result of an embryologic defect although its embryologic origin remains a subject of debate. When aberrant, however, the artery comes off the aortic arch beyond the left subclavian artery and crosses the midline, most often dorsal to

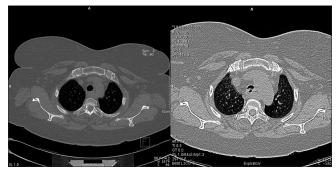


Figure 1: Axial lung section of the upper thorax in inspiratory phase (left) showing normal tracheal dimension and expiratory phase (right) showing more than 50% reduction of tracheal dimension suggesting tracheomalacia



Figure 2: Reformatted coronal image (left) and axial contrast-enhanced computed tomography (right) demonstrating aberrant right subclavian artery at the level of focal tracheomalacia



Figure 3: Volume rendering technique coronal image demonstrating aberrant right subclavian artery

the esophagus (80%), but may also pass between the esophagus and the trachea (15%), or even anterior to the trachea (5%).^[8]

Symptoms associated with aberrant right subclavian artery commonly involve the gastrointestinal and respiratory tracts. Symptoms result from the compressive

effect of the artery or aneurysm on the surrounding structures. Dysphagia is the most common and best-recognized symptom. When it compresses the tracheobronchial tree, respiratory symptoms are manifested. Cough in such cases has been attributed to the pressure effect exerted on the trachea by the esophagus. Such patients have both dysphagia and cough. Very rarely, it can present as a radiological abnormality mimicking mediastinal adenopathy in otherwise asymptomatic adults. [9] Isolated direct compression of the trachea by the aberrant vessel as is demonstrated here is an extremely rare occurrence.

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Conflicts of interest

There are no conflicts of interest.

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