The Expert Clinician

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A 37-Year-Old Woman with Dyspnea and Stridor

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In Brief

A woman with morbid obesity and a diagnosis of asthma since childhood sought medical attention because of unrelenting dyspnea and nocturnal stridor. Spirometry revealed a restrictive pattern with amputation of inspiratory and expiratory maximum flow rates. She was referred for further diagnostic evaluation.

Case Vignette

A 37-year-old morbidly obese woman presented with a history of asthma that developed in childhood. She reported recent progression of symptoms, now with dyspnea on limited activity and nocturnal stridor. A weight gain of 20 kg over 2 years was reported. Treatment with an inhaled corticosteroid and an inhaled long-acting β-agonist and intermittent administration of an oral or intravenous corticosteroid drug failed to improve her symptoms.

On physical examination, she was afebrile; her blood pressure was 135/88 mm Hg, pulse was 82 beats/min, respiratory rate was normal, and body mass index was 49 kg/m^2. Oxygen saturation was 95% while she breathed ambient air. The lungs were clear to auscultation, and the remainder of the physical examination was unremarkable.

Laboratory testing showed a normal hemoglobin, platelet count, and white blood cell count, without eosinophilia.

Table 1. Principal spirometric data of the patient

	Predicted	Before BD	% Predicted	After BD	% Predicted	% Change
FVC, L	3.16	2.34	74	2.5	79	7
FEV_1, L	2.72	1.85	68	1.98	73	7
FEV_1 /FVC, %	81.5	79.1	97	79.2	97	0
FIV_1, L	2.72	1.52	56	1.51	56	0
PEFR, L/s	6.49	2.55	39	2.42	37	−5
PIFR, L/s	6.49	1.44	22	1.48	23	3

Definition of abbreviations: BD = bronchodilator; FIV_1 = forced inspiratory volume in 1 second; PEFR = peak expiratory flow rate; PIFR = peak inspiratory flow rate.

Biochemistry was within normal ranges, and IgE level was normal. Aspartate aminotransferase, alanine aminotransferase, lactate dehydrogenase, C-reactive protein, and creatine kinase were within normal limits. Antinuclear antibodies, double-stranded DNA antibody, rheumatoid factor, complement levels, and anti–neutrophil cytoplasmic antibody tests were negative or in the normal range. The results of spirometry are shown in Table 1 and Figure 1. FEV_1/FVC before and after inhalation of a bronchodilator was 0.791 and 0.792 liters, respectively.

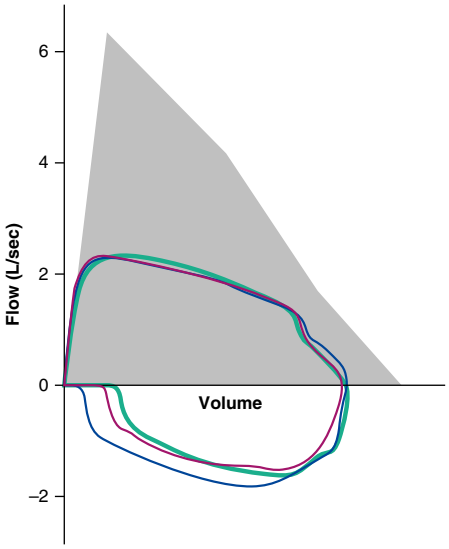

Figure 1. Spirometric flow–volume loop.

Question

1. What are the appropriate next diagnostic studies?

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Clinical Reasoning and Further Diagnostic Evaluation

Our patient presented with worsening dyspnea and the recent onset of stridor. Her symptoms did not improve with treatment for asthma. Other features of the presentation were also atypical for asthma. Symptoms typically include some combination of wheezing, dyspnea, chest tightness, and chronic cough. Asthma symptoms are generally episodic when adequate control is not achieved, and the symptoms are usually worse in the early hours of the morning. Asthma symptoms are often provoked in response to one or more identifiable triggers. None of these typical features of asthma in a younger adult were present in our patient. We considered asthma mimics such as granulomatous vasculitis, eosinophilic lung disease, parasitic infections, or an obstructing airway abnormality such as aspiration of a foreign body, tracheomalacia, central airway granulomas, or an endobronchial tumor. Examination of the spirometric flow–volume loop showed flattening of both the inspiratory and expiratory limbs compatible with a fixed obstruction of the trachea (Figure 1).

Chest computed tomographic (CT) imaging showed a reduction in the tracheal diameter throughout its length (6.7–6.3 mm), a trifurcated main carina, and normal caliber of the main bronchi (Figures 2A and 2B). Three-dimensional reconstruction allowed for observation of complete tracheal rings without a membranous posterior wall (Figure 3A). Repeat CT imaging after intravenous administration of a contrast agent revealed no associated intrathoracic vascular abnormalities. An echocardiogram was likewise normal.

After a multidisciplinary discussion, a flexible bronchoscopy was performed in a sitting position with a 5-mm-diameter bronchoscope. Local anesthesia and minimal sedation with midazolam and fentanyl was administered. There was immediate intolerance when entering into the trachea. After a rapidly obtained photograph (Figure 3B), the procedure was quickly interrupted, and the patient recovered.

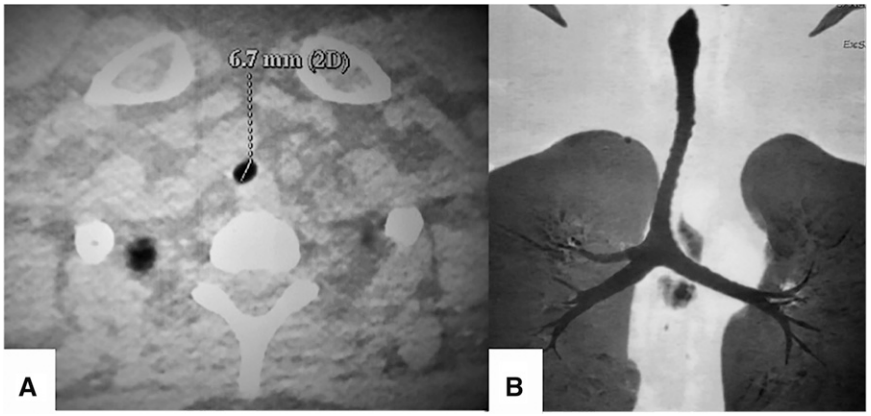

Figure 2. The computed tomography scan shows a tracheal constriction along the full tracheal length and a trifurcated main carina. (*A*) Transverse view of upper airway and trachea diameter. (*B*) Sagittal reconstruction of central airway and lungs.

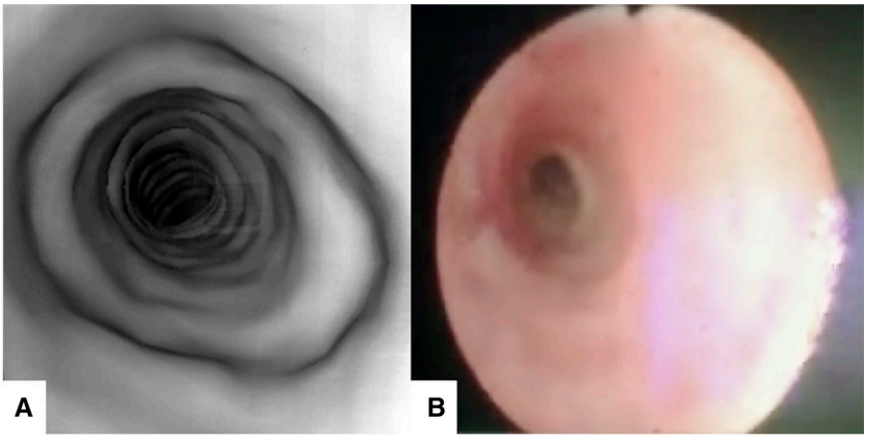

Figure 3. Three-dimensional tomographic reconstruction shows complete tracheal rings without a membranous posterior wall. Findings were confirmed by flexible bronchoscopy. (*A*) Three-dimensional reconstruction of tracheal rings. (*B*) Flexible bronchoscopic view of tracheal stenosis.

More Questions

2. What is the cause of the patient's dyspnea?

3. What other anatomic abnormalities are sometimes associated with this condition?

4. What is the appropriate treatment?

[Continue onto next page for answers]

Discussion

This constellation of anatomical and physiological findings encountered in our patient is diagnostic of congenital tracheal stenosis (CTS).

First discovery of this condition in adulthood is unusual but not unprecedented (1, 2). We believe that the patient's progressive dyspnea in the months leading to her presentation were caused by further airway narrowing associated with recent weight gain.

CTS, also called "stove pipe" trachea, is an uncommon disease. There are several classifications of CTS (1). Wolman and colleagues described two main types, A and B (2). Type A manifests limited tracheal involvement. In type B, tracheal involvement extends the entire length of the trachea and may compromise the main bronchi (2).

Subsequently, Speggiorin and colleagues (3) published a study of 84 pediatric patients with CTS and based classification on tracheal arborization. The authors found normal arborization in 62% of patients, tracheal bronchus of the right upper lobe in 11.9%, trifurcation (as in our patient) in 16.6%, and an airway to a single lung in 9.5%. Each of these may or may not be associated with bronchial compromise.

Treatment is dependent on symptoms, patient age, and airway anatomy. Asymptomatic patients are managed conservatively. For symptomatic patients with short tracheal stenoses, laser ablation of the posterior wall of the trachea, rigid bronchoscopic dilatation, or surgical tracheal reconstruction can be performed. Patients with a long segment of tracheal involvement may require a slide tracheoplasty as the best procedure (1). Other surgical treatments have been described. Rib cartilage augmentation, pericardial patch augmentation, and tracheal resection with end-to-end anastomosis can be performed (4). Because bronchomalacia has previously been described in association with CTS (4), an inspiratory and expiratory CT or full bronchoscopic evaluation should precede treatment.

In addition, there is frequent association with vascular defects that may require simultaneous repair. Atrial septal defect, coarctation of the aorta, patent foramen ovale, patent ductus arteriosus, pulmonary artery stenosis, and others have been described (5). CTS is seen in higher frequency in Down syndrome (1). Therefore, patients with CTS should be routinely evaluated for these genetic, congenital cardiac, and vascular anomalies.

The mortality associated with surgery is high (28% in one series). When CTS is seen with comorbid vascular anomalies, the untreated survival is less than 1 month (5, 6). Cheng and colleagues (7) reported a series of patients with mild symptoms and noncritical CTS who were treated conservatively. Mortality was lower than the group who received surgery (9 vs. 27%). Follow-up CT in the conservative treatment group (n = 6) showed that the trachea grew in all cases, and five of six had growth at a faster speed than normal, reaching near-normal size at age 9 years.

Few cases of CTS have been reported in adults, and all of them were incidental findings, managed with conservative management. We found eight reported cases in adults, including four patients during orotracheal intubation (8–12), one with repeated respiratory infections (13), another described in a tongue cancer study (14), and a final case described in an interstitial pneumonia workup (15). Of the eight published cases, four had a history of asthma.

Our patient was initiated on continuous positive airway pressure (CPAP) treatment due to clinical suspicion of sleep apnea, which was later excluded by polysomnography. She chose to continue overnight CPAP treatment because it provided partial relief of her nocturnal dyspnea, which may have been due to an increase in tracheal diameter and an associated improvement in work of breathing during sleep.

Answers

1. What are the appropriate next diagnostic studies?

CT imaging of the thorax and fiberoptic bronchoscopy.

2. What is the cause of the patient's dyspnea?

Congenital tracheal stenosis.

3. What other anatomic abnormalities are sometimes associated with this condition?

Additional anatomic abnormalities of the central airways, including circumferential tracheal rings, carinal defects, and tracheal or bronchial malacia; intrathoracic vascular anomalies.

4. What is the appropriate treatment?

Surgical interventions are considered for severely compromising stenosis presenting in infancy or childhood; most adults are treated conservatively.

Follow-Up

The patient was lost to follow-up for a period of 3 years. She later returned for consultation with the same symptoms accompanied by 10 kg of weight gain (body mass index, 53.5 kg/m^2) and no change in her "asthma."

Asthma treatment was withdrawn and she was continued on CPAP with 6 cm H_2O, even though polysomnography again did not show a sleep apnea association. Six months later, the patient reported good adaptation to CPAP without changes in spirometry.

Insights

- Congenital tracheal stenosis is a rare and potentially lethal malformation that is often associated with other anatomic anomalies of the central airways or great vessels.
- This disease generally manifests in the first months of life but rarely first comes to diagnosis after the age of 21 years.
- Congenital tracheal stenosis often requires surgical repair; however, for patients with mild symptoms, conservative treatment has been described. ■

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