Tracheobronchomalacia as a Rare Cause of Chronic Dyspnea in Adults

Meral Uyar^a Maruf Sanli^b Selim Kervancioglu^c Berna Taskin-Dogan^a
Kemal Bakir^d Oner Dikensoy^a

Departments of ^aPulmonary Medicine, ^bThoracic Surgery, ^cRadiology and ^dPathology, Gaziantep University, Gaziantep, Turkey

Keywords

Asthma · Amyloidosis · Bronchoscopy

Abstract

Objective: To emphasize the importance of a careful clinical evaluation to prevent unnecessary interventions and treatments. **Clinical Presentation and Intervention:** A 76-year-old female patient had been diagnosed with asthma during previous admissions to different hospitals. She had also undergone fiberoptic bronchoscopy (FOB) on 2 occasions for evaluation of right middle lobe atelectasis observed on computed tomography. A repeated FOB revealed tracheobronchomalacia and nodular bronchial amyloidosis. A silicone Y stent was inserted, but the dyspnea increased. Excessive granulation tissue developed, and the patient died despite ventilatory support. **Conclusion:** The stenting technique used did not prevent the development of respiratory failure and death in this patient. Hence, a surgical procedure could be considered as an alternative to stenting in such cases.

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Introduction

Tracheobronchomalacia (TBM) is characterized by tracheobronchial collapse during the exhalatory phase of respiration due to weakness of the airway walls [1]. The primary form of TBM occurs due to congenital absence of tracheal cartilage, mostly in infants [1]. In adults, a secondary form is seen most commonly in middle and older age groups. The secondary form of TBM may be caused by recurrent airway irritation, such as occurs with tracheobronchitis, smoking, chronic obstructive pulmonary disease, asthma, and connective tissue disorders, or it may be caused by mechanical factors, such as extrinsic compression, trauma, and intubation [1–3]. Tracheomalacia and TBM must be considered in cases of difficult-to-treat asthma, foreign-body aspiration, and endobronchial obstruction [1–3].

Tracheobronchial amyloidosis is a rare entity and it is slightly more common in females than in males [4]. Tracheobronchial amyloidosis occurs in two different forms: a diffuse submucosal form and a local nodular form [4].

Fig. 1. CT scan showing atelectasis of the right middle lobe.

Fig. 2. Bronchoscopic view demonstrating the total collapse of the trachea during expiration.

Tracheobronchial amyloidosis may be symptomatic or asymptomatic due to a secondary obstruction [4]. Here, we present a symptomatic case with TBM and nodular endobronchial amyloidosis.

Case Report

A 76-year-old female was admitted to our hospital with complaints of cough, dyspnea, night sweats, and weight loss. She reported cough and progressive dyspnea for the preceding 10 years. She had been diagnosed with asthma previously and had been on long-term home oxygen therapy due to chronic respiratory failure. Her medical history was remarkable for frequent emergency room visits in the 2 years preceding this admission. She had undergone percutaneous coronary intervention for coronary artery disease 5 years ago. She also had a history of diabetes mellitus, hypertension, and peptic ulcer disease. The patient had been admitted to another hospital 2 years prior for pneumonia. Fiberoptic bronchoscopy (FOB) was performed during that hospitalization for delayed resolution of pneumonia. Biopsy samples were nondiagnostic. FOB was repeated 2 months later at a different hospital. There was no report of excessive airway collapse during exhalation in the reports of these procedures. A careful family history revealed that her mother and sister died of respiratory failure, while her father died of heart failure. The physical examination was notable for bilateral rhonchi. Laboratory findings upon admission were as follows: white blood cell count, 9,000/mm³; hemoglobin, 14.6 g/dL; glucose, 162 mg/dL; blood urea nitrogen, 30 mg/dL; creatinine, 0.97 mg/dL; erythrocyte sedimentation rate, 11 mm/h; and C-reactive protein, 3 mg/L. Pulmonary function tests were as follows: forced vital capacity, 1.89 L (117% predicted); forced expiratory volume

in the first second, 0.99 L (77% predicted, with 10% reversibility); and forced expiratory volume in the first second/forced vital capacity, 52%. Computed tomography (CT) of the chest revealed right middle lobe atelectasis (Fig. 1). FOB was repeated to further evaluate the chest CT scan findings. There was a total collapse of the trachea and bilateral main stem bronchi during exhalation in bronchoscopy (Fig. 2). A white mucosal lesion obstructing the right middle lobe was observed. Histopathological examination of the biopsy samples was compatible with amyloidosis. Sputum and bronchoalveolar lavage sample results were benign. Specifically, they were negative for acid-fast bacilli and no growth was observed on microbial cultures. On the basis of these findings, TBM and coexistent nodular bronchial amyloidosis were diagnosed. A silicone stent (Dumont Y stent; France) was placed for the treatment of TBM. Ten days later the patient was readmitted due to progressive dyspnea. On that occasion she was admitted to the intensive care unit. Despite the use of noninvasive ventilation for insufficient ventilation, she ultimately required intubation. Bronchoscopic evaluation revealed granulation tissue in the distal portion of the stent. The stent was subsequently removed by rigid bronchoscopy under general anesthesia, and a repeated FOB was performed to relieve crusts. Despite utilization of high pressures with mechanical ventilation, target tidal volumes were not achieved. The patient died 1 week after hospitalization.

Discussion

The patient was diagnosed with TBM on FOB. TBM is diagnosed when the tracheobronchial lumen diameter decreases by 50% or more during exhalation. The diagno-

sis of TBM is made either by direct observation of collapse of the tracheobronchial system during FOB or by observation of a tracheobronchial collapse during exhalation on a dynamic thoracic CT scan [5]. Clinicians must consider TBM in the differential diagnosis of chronic dyspnea. The severity of TBM can range from mild to severe [6]. It is frequently misdiagnosed as asthma or chronic obstructive pulmonary disease as the clinical presentations are similar and TBM is a rare disease. Placement of a silicon stent is recommended for the treatment of severe cases of TBM. Rarely, complications, such as granulation formation, migration, and mucus plugs, may occur following stent placement [7, 8]. Unfortunately, the development of severe granulation tissue in the presented case resulted in a fatal airway obstruction despite aggressive

care and multiple interventions. An external stabilization technique is defined in TBM especially in children. This technique could be considered for appropriate patients [9, 10]. Surgical alternatives to stenting should be considered in patients with complications due to tracheal stenting.

Conclusion

The stenting technique used did not prevent the development of respiratory failure and death in this patient. Hence, a surgical procedure could be considered as an alternative to stenting in such cases.

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