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CASE REPORT

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Unexplained disabling and long-lasting cough: a case report

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Abstract

Background A 51-year-old woman was referred to our department due to chronic dry cough lasting six years without an etiological diagnosis. The patient suffered from chronic deterioration in her quality of life due to a persistent cough that sounded like a barking seal.

Case presentation A severe form of malacia involving the inferior third of trachea and the main bronchi was diagnosed. According to our protocol, a self-expandable prothesis was placed in the distal portion of the trachea via rigid bronchoscopy with excellent results in cough relief. The patient was subsequently scheduled for tracheobronchoplastic surgery with a polypropylene matrix. Two and a half years after surgery the patient had a significant improvement in quality of life with a complete resolution of her symptoms.

Conclusion This report demonstrated that tracheobronchomalacia can be difficult to diagnose with a serious impact on the patient's life. Referral to a specialized center is essential in the diagnostic and therapeutic management of this disease. Surgical treatment by tracheobronchoplasty may represent a good solution in selected patients.

Keywords Tracheomalacia, Tracheobronchoplasty, Tracheobronchomalacia, Rigid bronchoscopy, EDAC

Introduction

Tracheomalacia is a rare pathology characterized by congenital or acquired structural abnormality of the trachea. When the disease extends to mainstem bronchi, it is identified as tracheobronchomalacia (TBM). The diagnosis may be difficult because of many nonspecific symptoms like dyspnea and cough, that could be correlated to other common respiratory diseases such as COPD. Diagnosis is confirmed by dynamic tests: flexible bronchoscopy and/or computed tomographic (CT) scan of the chest. Some management algorithms have been proposed, but no specific recommendation was established. In this article, we present a patient with long-lasting debilitating cough who, after years of struggling, got a TBM diagnosis and the appropriate treatment.

Case presentation

A 51-year-old woman was referred to our department after multiple medical assessment for dry chronic cough persisting for six years. Her medical history began with recurrent bronchitis and otitis, mumps meningitis and whopping cough at 5 years of age. At the age of 7, she tested positive for Mantoux intradermal reaction, but no contact with Mycobacterium tuberculosis was identified.

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Other comorbidities included a hypofunctioning goiter, psoriasis, endometriosis, gastroesophageal reflux disease (GERD) and Penicillin dermal reaction. Ultrasound thyroid examination ruled out local compression of the airway. An esophagogastric endoscopy showed a small hiatal hernia and mild gastric and esophageal inflammation with histological aspects of subacute-chronic esophagitis. The pH esophageal manometry showed abnormal acid reflux and ineffective esophageal motility. Four years earlier, she had a bronchoscopic evaluation in another institution, reported as normal.

Her quality of life and social relationships were severely impacted by symptoms. She reported difficulty sleeping due to the persistent cough that led to mental and physical exhaustion. Her social life was greatly affected especially in the Covid-19 pandemic context, where coughing became a reason for interpersonal distancing and discrimination.

At the time of examination, the patient had a body mass index (BMI) of 29,3 and she reported for six years, a chronic, invalidating cough that sounded like a barking seal. Until then she had received proton-pump inhibitors, antitussive drugs and aerosol therapy.

Respiratory function tests did not show obstruction, no bronchial hyper-reactivity at methacholine test; allergic and auto-immune etiologies were explored without any pathological findings.

A new evaluation by fiberoptic bronchoscopy was performed in our hospital showing a severe malacia involving the inferior third of trachea, more prominent on the left side, and the main bronchi (Video). A decrease of 90% of the airway diameter was observed during forced expiration. Cytological and bacterial sampling (including tuberculosis) was negative. Random endoscopic biopsies of the airway showed only a mild inflammatory status.

A dynamic CT revealed a significant decrease of the tracheal diameter during forced expiration and multiple nodules in the lung parenchyma. A diagnosis of diffuse idiopathic pulmonary neuroendocrine cells hyperplasia (DIPNECH) was suspected. The largest of these nodules (7 mm in the left lower lobe) was removed by video-assisted thoracoscopy and the histological report showed no abnormalities, so DIPNECH was excluded.

The patient was diagnosed with TBM and, after discussion and validation by a multidisciplinary team, was scheduled for placement of a tracheal prosthesis under rigid bronchoscopy. In 2018 our centre started a pilot study based on the experience of Buitrago et al.[1] and his team in Boston, consisting in surgical stabilization of the central malacic airways by posterior splinting with a prosthetic mesh (tracheobronchoplasty, TBP) after a first stent-trial step.

A self-expandable 40×20 mm Silmet metallic stent was placed in the distal portion of the trachea (Fig. 1) via rigid bronchoscopy with excellent results in cough relief, without postprocedural complications.

The patient was then scheduled for stent removal after two weeks and selected for TBP. This surgical technique, as described by Grillo [2], consisted in the positioning of a polypropylene matrix sutured to the back wall of the trachea and mainstem bronchi via right posterolateral thoracotomy in the fourth intercostal space (Fig. 2). Postoperative course was uneventful with complete resolution of her symptoms. After two and a half years the patient still reported a significant improvement in quality of life with complete disappearance of coughing

Discussion

The diagnosis of this case of TBM was greatly delayed with respect to the onset of symptoms, to the detriment of the patient's quality of life. There is currently no consensus on the diagnostic criteria for large airway collapse (LAC) or Expiratory Central Airway Collapse (ECAC), so it can be object to subjective interpretation. ECAC is traditionally defined as 50% modification in the tracheabronchial diameter during breathing and it is present in one out of three patients with chronic obstructive pulmonary disease (COPD) or severe asthma [3]. Two types of ECAC were described: Excessive Dynamic Airway Collapse (EDAC) with an inward bulging of the atrophic muscular fibers in the posterior airway membrane during exhalation with narrowing of the cross-sectional airway lumen; and TBM characterized by weakness of the anterior tracheobronchial cartilage wall with or without excessive dynamic invagination of the posterior membranous wall [1]

Wright and Mathisen [2] identified a threshold of 90% in airway collapse, combined with severe malacia with exhalation during quiet breathing to validate TBM diagnosis. There are different forms of tracheobronchomalacia in adults: primary (genetic, idiopathic) or secondary to trauma, tracheotomy, intubation, surgery, transplantation, emphysema, infection, inflammation, chronic bronchitis, extrinsic compression; or undiagnosed in childhood vascular rings [4]. Dynamic thoracic computed tomography (CT) and fiberoptic bronchoscopy with spontaneous breathing patient are essential for diagnosis; these investigations allow to evaluate the behavior of airways during inhalation and exhalation and the extent of the disease [5]

Our case was probably an acquired TBM whose cause remains unclear. Tracheal compression, COPD and inflammatory causes were excluded during hospitalization. Prolonged intubation and trauma lack in the medical history while recurrent bronchitis experienced in pediatric age could explain TBM. Moreover, this patient has GERD in her medical history that can be associated with TBM [6]. It is unclear whether GERD is a cause or a

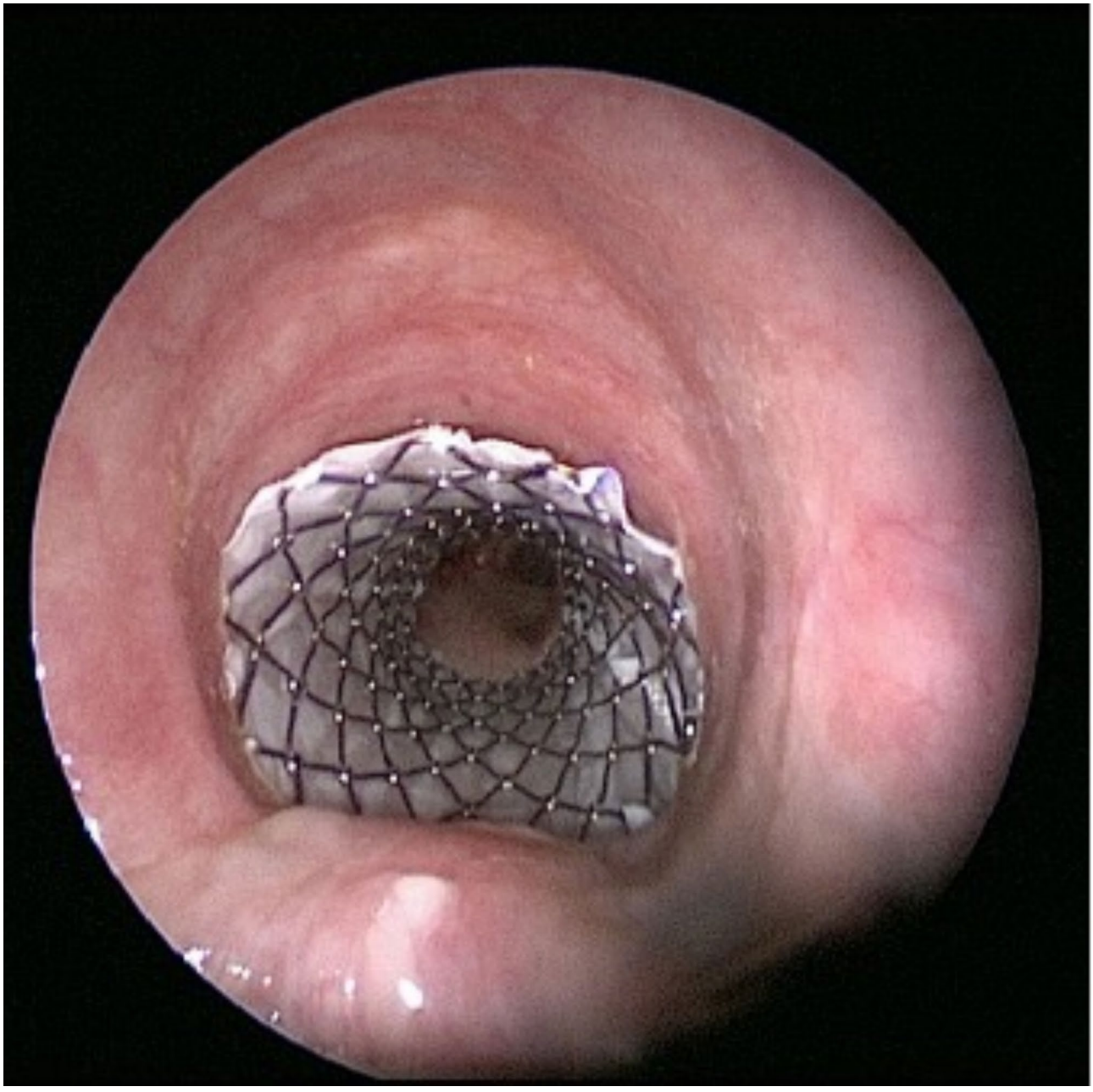

Fig. 1 bronchoscopic view of metallic stent in the lower trachea

consequence of TBM, even if a pathogenetic mechanism has been proposed for pediatric population [7]

TBM is also associated with inhaled corticosteroids with a stronger association for higher doses and longer use but a cause-effect relationship is far to be proved, because of study limitations [7]

However, in our case, the diagnosis of TBM took almost 6 years. The previous fiberoptic bronchoscopy was performed in another institution and exam video recording was not available. An extremely late onset of congenital TBM cannot be ruled out with certainty, even if the airway was described as normal

Airway stenting [2, 5] could be an effective option for both surgical candidates and nonsurgical patients. Hence, airway stenting works as a trial for symptom improvement before TBP because it simulates tracheal and/or bronchial stabilization. Wright and Mathisen [2] bring to attention that some patients with TBM could not tolerate stent positioning because of airway irritation with cough persistence or amplification, so this solution is not always possible. According to Buitrago et al.[1], a stent

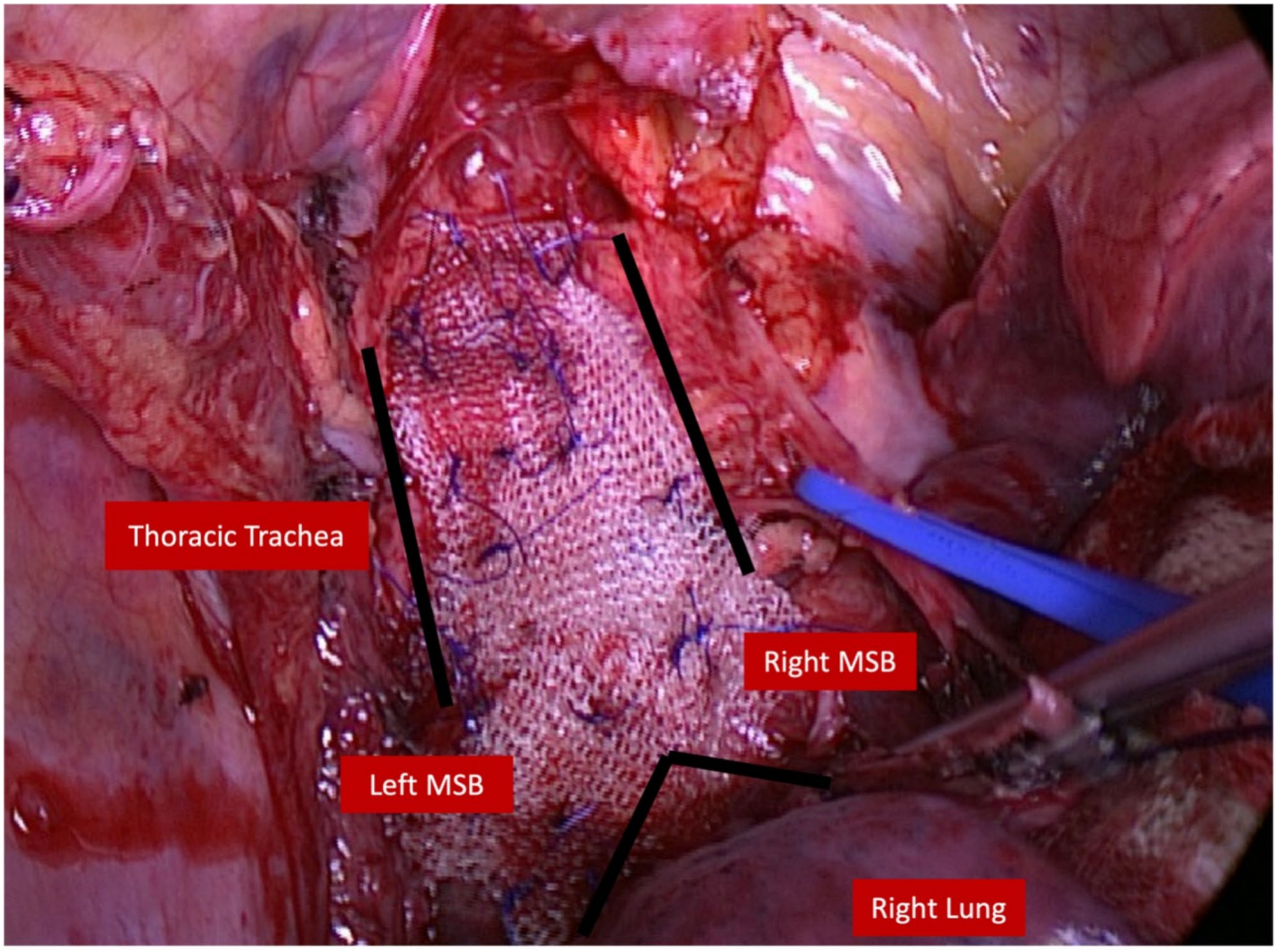

Fig. 2 Intraoperative view of polypropylene non absorbable mesh sutured on the thoracic portion of trachea and main bronchi. MSB: mainstem bronchus

was firstly placed. This strategy allows the best selection of patients with EDAC and TBM who may have the best response to TBP. On the other hand, TBP is an effective option independently by the type of malacia (anterior or posterior) because the polypropylene mesh stabilizes the posterior wall avoiding bulging into the lumen and keeps the distance between cartilaginous rings fixed, giving stability to anterior wall of trachea [2]. To date, tracheal replacement is not yet indicated in TBM [8, 9]

Conclusion

TBM is a rare disease with nonspecific symptoms, common to most respiratory pathologies. Overlap with other respiratory diseases is common (e.g. COPD, airway involvement of inflammatory or autoimmune diseases) and this contributes to the complexity of the diagnosis. This report provides a perfect example of non-overt disease presentation and difficulty in identifying TBM. Referral to a specialized center is essential for the diagnosis and therapeutic management of this disease to propose the most appropriate treatment. Tracheobronchoplasty may represent a good solution in selected patients to restore a normal configuration of the airway

Abbreviations

BMI	Body mass index
COPD	Chronic obstructive pulmonary disease
CT	Computed tomographic
DIPNECH	Diffuse idiopathic pulmonary neuroendocrine cells hyperplasia
ECAC	Expiratory central airway collapse
EDAC	Excessive dynamic airway collapse
GERD	Gastroesophageal reflux disease
LAC	Large airway collap
TBM	Tracheobronchomalacia

Supplementary Information

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Supplementary Material 1

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Not applicable

Consent for publication
The patient provided her written informed consent to participate in this study and to publish the video.

Competing interests
The authors declare no competing interests.

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