Tracheobronchomalacia and/or Excessive Dynamic Airway Collapse?

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ABSTRACT

Expiratory airway collapse is defined as more than 50% reduction in the horizontal cross-sectional area of the airway at expiration or during coughing; and is due to pure tracheobronchomalacia (TBM) or excessive dynamic airway collapse (EDAC). It is not easy to make a clear differentiation between TBM and EDAC. These two conditions are different entities, yet they may be mistaken for each other and they may also coexist. In this paper we discuss differential diagnosis of EDAC and TBM in the light of fiberoptic and virtual bronchoscopic images according to a newly proposed FEMOS classification.

Key Words: Tracheobronchomalacia, tracheomalacia, bronchomalacia

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Introduction

Expiratory airway collapse is defined as more than 50% reduction of the horizontal cross-sectional area at expiration or during coughing and is due to pure tracheobronchomalacia (TBM) or excessive dynamic airway collapse (EDAC) (1). It is not easy to make a clear differentiation between TBM and EDAC. These two conditions are different entities, yet they may be mistaken for each other and they may also coexist. In this article, pathological airway collapse depicted in two cases are discussed with reference to the diagnoses of TBM and EDAC, in the light of the recently proposed FEMOS classification (2).

TBM is defined as the flaccidity of tracheal and bronchial walls due to softening of cartilaginous support or to hypotonicity of the myoelastic structure. The trachea and main bronchi lose their normal stiffness and airway walls come close to each other in expiration. The clinical picture consists of dyspnea, difficulty in expelling secretions, coughing, wheezing or pneumonia (3). TBM is being diagnosed and treated more frequently, but it is thought that the condition is still underdiagnosed. Also, there is ambiguity regarding the appropriate therapy (4).

EDAC is defined as 50% or more reduction of the sagittal diameter of the trachea in forced expiration or during coughing. Primarily, it is characterized by an excessive collapse of the posterior membranous trachea towards the lumen. The tracheal cartilages are structurally and functionally normal in EDAC, and EDAC has a better prognosis than TBM (3).

The word FEMOS is an acronym for the words ‘function’, ‘extent of abnormality’, ‘morphology’, ‘origin of disease’ and ‘severity of airway collapse’. This classification is proposed in order to delineate a common language for the definition of diseases that have a major dynamic airway collapse, and also for clinical reasons, which include identifying the benefits of treatment (2).

Case 1: A 63 year old, ex-smoker male with chronic shortness of breath. There was pulmonary tuberculosis and thoracic trauma in his history. He was being hospitalized approximately 2-3 times a year due to exacerbations of COPD. His best PFT values, acquired while he was not in an exacerbation phase, were: FVC: 1.13 L (29%), FEV1: 0.55 L (18%), FEV1/FVC: 53%; and the arterial blood analysis (ABG) read as: with 2 L/min oxygen, pH: 7.39, pO2: 70.4 mmHg, pCO2: 54.8 mmHg, HCO3: 32.9 mmol/L. The chest x-ray and thorax CT indicated emphysematous areas in both lungs. During fiberoptic bronchoscopy (FOB), the posterior membranous trachea showed intraluminary collapse throughout all its segments, to such an extent that the trachea touched the anterior wall during expiration and coughing, and that this condition continued to the two main bronchi and most of the lobar bronchi. The anterior and lateral tracheal walls and the tracheal cartilages were observed to be normal (Figure 1). When evaluated according to the FEMOS classification, the case was classified as having crescent EDAC due to COPD and Mounier-Kuhn syndrome, with a functional status of F4, disease extent of E4, and severity of airway collapse of S4. Surgical consultation and non-invasive mechanical ventilation options were tried but no additional treatment to home oxygen and medical treatment could be given. The patient died due to a COPD exacerbation after two years of this evaluation.

Case 2: 59 year old male with COPD for 7 years had exacerbations that required hospitalization approximately 2-3 times a year. During optimal clinical status, his PFT was as
follows: FVC: 1.04 L (26%), FEV1: 0.55 L (18%), FEV1/FVC: 53%, and ABG analysis was pH: 7.43, pCO2: 42.6 mmHg, pO2: 90 mmHg, Sat: 97.2%. In thoracic CT, extensive hyperinflation and chronic changes with subsegmental atelectasis in the lateral basal segment of the lower left lobe were observed. During FOB, luminal narrowing was observed in all tracheal walls, with a sabre-sheath morphology in the middle and upper 1/3 parts of the trachea, and there was an 80% reduction of the intratracheal lumen during forced expiration (Figure 2). The images from the lower end of the trachea and both main bronchi were different from those of the above, with the tracheal antero-posterior diameter reducing up to 90% during expiration, while the anterior and lateral walls of the trachea were almost normal (Figure 3). When evaluated by the FEMOS classification, the etiology was COPD; morphology was sabre-sheath at some locations, circumferential at some other locations and crescent at others. The functional status was F3, extent of disease was E3 and severity was S3. The disease had features of both focal TBM and multifocal EDAC. The dynamic and virtual CT bronchoscopic images were in accord with FOB (Figure 4). Surgical consultation and non-invasive mechanical ventilation options were tried, but no additional treatment to home oxygen and medical treatment could be given. The patient could be followed for three years and presumed to be alive.

Discussion

When we investigated the two cases in detail, we evaluated the first case as EDAC and the second case as TBM + EDAC. The first case fits precisely to the definition of EDAC, and thus the FEMOS classification could appropriately be applied to this case. However, since TBM was not diffuse and there were areas compatible with EDAC at other sites, the second case led us to consider that the FEMOS classification needed to be elaborated regarding diagnosis.

In this article, we aimed to emphasize the importance of differentiating dynamic airway collapse (DAC) from TM. However, it is known that these two entities may coexist, which was the case in our second patient. The coexistence of these two diagnoses originates from a similar physiopathologic condition (3). We know that, normally, the diameter of the trachea is reduced only less than 50%, during coughing. This normal collapse may be somewhat exaggerated in severe asthma and emphysema, due to changes in airflow velocity through the airways. The increase in airflow in a focal area results in an increase in transmural pressure, which leads to airway collapse. Some studies have indicated that the airway collapse in COPD is secondary, or a later developed TM. It has been reported that, contrary to primary TM patients, no significant progression was observed in these cases (1).

A review of the literature indicates that the cases undergoing bronchoscopy have a 4-23% overall diagnosis of EDAC or TBM (2, 3). Acquired TBM and EDAC may arise due to primary or secondary reasons. Secondary reasons are reported as long-term intubation and mechanical ventilation; undiagnosed tracheal cartilage fractures following closed chest traumas, especially steering-wheel injuries; chronic inflammations like relapsing polychondritis; tracheal cartilage damage due...
to tumors; postsurgical mechanical reasons like the postpneumectomy syndrome; chronic infections like tuberculosis; chronic compression due to substernal goiter, mediastinal tumors or vascular aneurysms; Mounier-Kuhn syndrome; Ehler-Danlos syndrome; thyroid diseases; electrocauterization, especially if applied longer than 3 seconds; asthma; and COPD (1). Particularly, asthma and COPD are reported as coexisting with EDAC (3). Both our cases were COPD with an asthmatic component.

EDAC is diagnosed with FOB and dynamic and virtual CT or cineMRI (Magnetic resonance imaging) (5, 6). In bronchoscopic inspection, the integrity of the patient's airways and the tracheal cartilage is evaluated in real-time during coughing and valsalva maneuvers. In both cases, the dynamic and virtual bronchoscopic images were in accord with bronchoscopic findings. The CT tracheobronchscopy, when compared to bronchoscopy, has the advantage of displaying the neighboring structures to the bronchial tree, and thus depicting a probable extrinsic compression. Being a noninvasive method is another of its advantages; it might be utilized in patients with respiratory distress or those that do not want to undergo FOB. In patients diagnosed with TM, CT reconstructions may be utilized for deciding a therapeutic procedure such as stenting. It may also be useful after treatment, in evaluating airway patency or stent complications. In spite of these advantages, multidetector CT is only complementary to fiberoptic bronchoscopy. The forthcoming progress seems to be towards acquiring real time bronchoscopic studies with CTs (7).

Various inflammations which proceed with diffuse airway narrowing, such as polichondritis, ulcerative colitis, amiloidosis, sarcoidosis, Wegener's granulomatosis, tracheopathia osteochondroplastica and papillomatosis may also present with similar symptoms and clinical findings. Dynamic CT is also useful in the differential diagnosis of these diseases (4, 8).

Treatment might be medical or invasive. Medical treatment encompasses treatment of the underlying disease and noninvasive mechanical ventilation. Acquired TBM and EDAC patients can notably benefit from CPAP >6 cmH2O. In both of our cases, while hospitalized, we have tried CPAP and BIPAP, but the patients could not tolerate these treatments due to increased hyperinflation. Airway stents are being tried often as minimal invasive therapy. However, it has been reported that, because the malacic choke point slides below the stent, more than one stent might be required and this will bring about a problem of increased secretions. Since, due to limitations in airflow, collapse occurs in smaller airways in COPD patients, tracheal stenting is not considered in patients with DAC. Also, the FDA has recently published an alert that metal stents not to be used in benign airway diseases (4). More invasive surgical interventions are generally considered only as experimental treatments (9).

In conclusion, new treatment options are needed for these patients and, although the recently proposed FEMOS classification has made a significant clarification regarding functional status, extent and severity of the disease, morphologically a clearer definition is needed.

Conflict of Interest
No conflict of interest was declared by the authors.

References