

Bronchial stenosis secondary to epidermolysis bullosa successfully treated with bronchoscopic balloon dilatation

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Abstract

Epidermolysis bullosa rarely affects lower airways. We present a case of lower airway involvement and stenosis successfully managed with flexible bronchoscopy and balloon dilation.

Case Report

A 19-year-old male patient with a history of epidermolysis bullosa (EB) and tracheostomy performed before he was 1 year old was referred to our interventional pulmonary service (July 2012) by his pulmonologist for progressive dyspnea, wheezing, mild hemoptysis, and abnormal computer tomography (CT) scan of the chest (Fig. 1). The patient was reported to have tracheal, upper and lower airways involvement with chronic sloughing, inflammation, and obstruction documented on previously performed diagnostic bronchoscopies. Prior to his referral, a chest roentgenogram done at the time of initial evaluation showed left lower lobe atelectasis. The patient was started on bronchodilator therapy for suspected asthma without any improvement (pulmonary function testing could not be performed). The hemoptysis was thought to be from his underlying mucosal disease and airway injury from the tracheostomy tube. As bronchodilator therapy did not improve

his symptoms, his Shiley tracheostomy #4 was upsized to a #6 Shiley with improvement in wheezing, but not the dyspnea. He underwent diagnostic bronchoscopy for definitive diagnosis, which revealed a tracheal and bronchial mucosa that appeared desquamated, erythematous, and friable in all areas. The left main stem bronchus appeared completely occluded with inflammatory tissue revealing a pinhole lumen. The bronchoscope could not be passed beyond the obstruction, even when this was attempted with a pediatric bronchoscope. The patient proceeded to have therapeutic flexible bronchoscopy (BF-1T180 large bronchoscope whose distal end measured 6 mm with a 3-mm working channel) in the operating room under general anesthesia. After establishing adequate sedation, the tracheostomy tube was removed as it would not accommodate the therapeutic bronchoscope. The bronchoscope was inserted through the tracheostomy stoma to access the trachea. Extra care was taken to avoid trauma to the airway mucosa during cannulation and de-cannulation

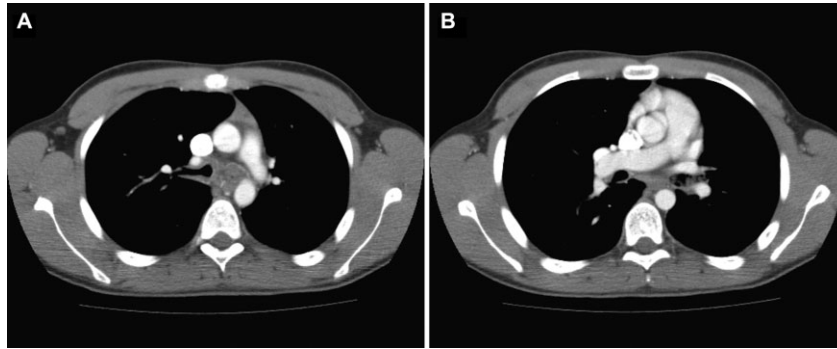


Figure 1. (A) Complete occlusion of the left main stem bronchus with intraluminal debris. (B) Narrowed, but patent lumen distal to the obstruction.

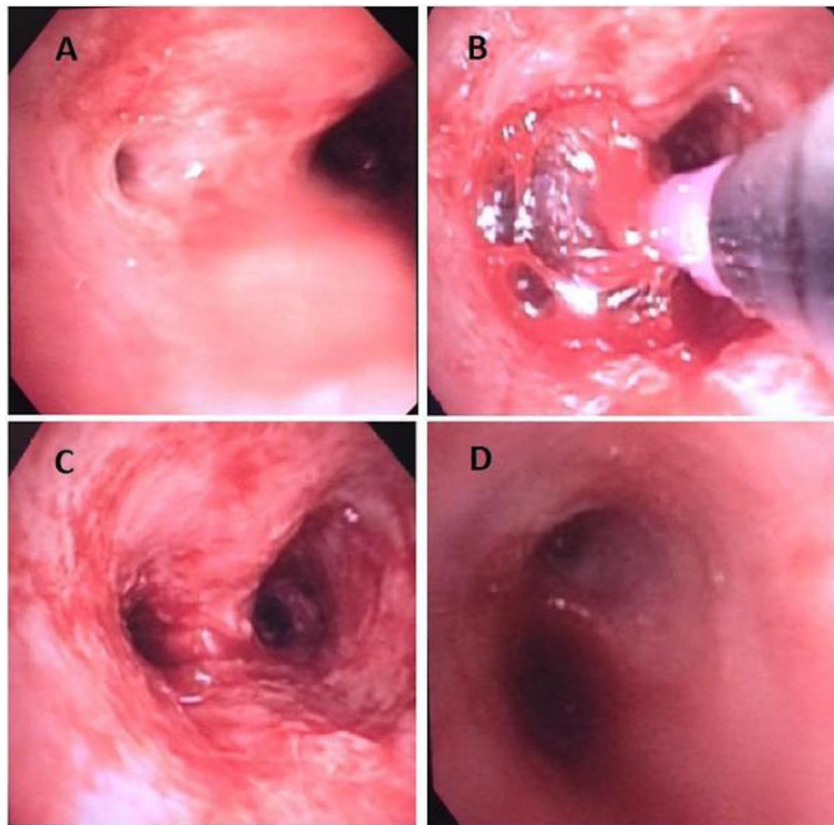


Figure 2. (A) Severe narrowing of the left main bronchus. (B) Balloon dilatation of the left main stem bronchus. (C) Significant improvement in the lumen diameter of the left main stem bronchus. (D) Visualization of the left upper and lower lobe bronchi post dilatation.

of the trachea when respiratory support was provided. Multiple balloon dilatations (inflation up to 12-mm diameter) of the left main stem bronchus were performed through the working channel of the bronchoscope (Fig. 2B). Significant lumen patency was restored, up to 80% of the normal diameter (Fig. 2C). At the end of the procedure, the left upper lobe and left lower lobe

bronchi were easily visualized (Fig. 2D). Repeat diagnostic bronchoscopy 3 months after the left main stem bronchus dilatation revealed continued patency of the airway. The patient reported significant improvement of his dyspnea. He required less bronchodilator use and experienced improved exercise capacity. He did, however, present 6 months after initial dilation with a similar degree of

obstruction and dyspnea. Similar flexible bronchoscopy was performed under similar settings with balloon dilation again leading to 80% patency of the left main bronchus lumen. He continues to be seen regularly by his pulmonologist, his last repeat CT scan from December 2013 showed persistent patency of his left main bronchus and has not required repeated therapeutic bronchoscopies since.

Discussion

EB is a group of mucocutaneous diseases that result from mutations in genes that code for structural proteins that maintain dermal integrity. Extra cutaneous manifestations can include involvement of nails, hair, teeth, ocular, oropharyngeal, laryngeal, gastrointestinal, urogenital, musculoskeletal, and cardiovascular processes.

Several case reports have described laryngotracheal involvement with symptoms ranging from increased secretions, recurrent bullae, fibrin accumulation, laryngeal, subglottic and tracheal desquamative disease, and de novo cicatricial lesions leading to stenosis. All these lesions lead down a common pathway of airway compromise as a life-threatening complication [1]. Airway involvement typically leads to laryngeal and tracheal stenosis with a cumulative incidence up to 40% by age 6. Many of these patients require tracheostomy or laryngeal interventions at an early age, just like our patient. The inclination to avoid endoscopic procedures in patients with EB was based on the risk of iatrogenic injury, poor wound healing, and the already poor prognosis of the disease [1, 2]. Previous interventions to improve airway compromise were done at the level of the larynx and the supra-glottic region. In a series of eight patients by Ida *et al.*, five patients underwent laryngeal surgical intervention and did not sustain any iatrogenic injuries, showed good wound healing, and did not require any repeated procedures [1]. Seven of those patients were found to have laryngeal pathology during rigid endoscopy, but there is no mention of downward involvement of the trachea or lower airways. In a case report by Gonzalez *et al.*, the authors suggest that previous reports have been made of patients with both tracheal and bronchial involvement [3]. Overall involvement of the laryngeal and tracheal mucosa is seen in very severe forms of the disease.

Despite tracheostomy and supportive measures, the mortality remains very high and the need for chronic tracheostomy is common. These patients often require frequent catheter suctioning and respiratory tract washing to temporarily clear the airways of increased secretions and

“sloughed mucosa.” The constant manipulation of these already vulnerable airways results in stenotic lesions, increased granulation debris formation and hemorrhages. To our knowledge, intrathoracic and lower airway dilation or stent has not been reported, despite the obstructive nature of the disease [2].

We initially hesitated to perform therapeutic bronchoscopy on this patient. The nature of the disease and the potential friability of these airways were the two main concerns. Several technical aspects are important to discuss in this case. First, we intentionally avoided rigid bronchoscopy because of obvious risks of airway tears and severe injuries. Second, we exercised extreme caution in cannulating and de-cannulating his trachea during respiratory support or interruptions of his therapeutic bronchoscopy. We were concerned that even small mucosal tracheal injuries may cause significant bleeding, which may have led to airway compromise and interference with bronchoscopic visual field. Third, we elected only to gently and incrementally dilate with an airway balloon, while avoiding rigid or thermal injuries such as forceps debulking or laser ablation. One last point, recurrences are expected in this group of patients, considering the nature of the disease. We believe with careful planning and execution, the goal of safely improving symptoms with therapeutic flexible bronchoscopy could be achieved. The main question remains about the longevity of these interventions, but we also believe the benefits outweigh the risks ratio in this group of patients who carry significant comorbidities and high surgical risks.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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