Severe bilateral bronchial stenosis with acute respiratory failure from granulomatosis with polyangiitis

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Introduction
Granulomatosis with polyangiitis (GPA), formerly named Wegener’s granulomatosis, is a small-vessel vasculitis associated with proteinase-3 anti-neutrophil cytoplasmic antibodies (PR3- or C-ANCA). GPA affects multiple organs, mainly the urinary and respiratory systems. Approximately 15–55% of GPA patients have tracheobronchial manifestations, which often lead to misdiagnosis with other common obstructive airway diseases such as asthma [1]. In this report, we present a 48-year-old woman with severe uncommon respiratory complications of GPA.

Case Report
In a 48-year old female, diagnosis of GPA was made by histological finding of granulomatous inflammation from nasopharyngeal biopsy and positive PR3-ANCA from blood test. In December 2014, her kidneys became involved. The kidney biopsy showed crescentic glomerulonephritis. Treatment with 50 mg daily of oral cyclophosphamide and 50 mg (1 mg/kg/day) of prednisolone was started, and the dosage of prednisolone was slowly tapered to 20 mg/day within 6 weeks.

In late January 2015, the patient came to our emergency department (ED) with progressive dyspnoea, wheezing, and respiratory failure due to bilateral main bronchial stenosis from granulomatosis with polyangiitis (GPA) involvement. By computed tomography imaging and flexible bronchoscopy, we measured the narrowest diameter at 2 mm. The patient promptly recovered from respiratory failure after treatment with flexible bronchoscopic balloon dilatation (BBD) without any procedure-related adverse event. This report showed the benefits of urgent flexible BBD that was used as a rescue therapy in a GPA patient who presented life-threatening acute respiratory failure from severe bilateral bronchial stenosis.
A flexible fiberoptic bronchoscope (model BF-P150; Olympus Medical System Corp., Tokyo, Japan) was passed through the tracheal tube. A flexible guide wire (Safe-T-J curved, 180-cm long; Cook Inc., Indiana, USA) was inserted through the working channel of the flexible bronchoscope and advanced through the stenotic segment of the bronchus. Then the bronchoscope was withdrawn while the guide wire was left in place. Next, the ventilator circuit was briefly disconnected from the tracheal tube before passing the balloon catheter (PTA catheter: Armada 35; Abbott Laboratories Vascular Enterprises, Beringen, Switzerland) over the guide wire under direct bronchoscopic visualization. The balloon chosen in this case was 10 × 40 mm to ensure 1 cm beyond each end of the stenotic segment to prevent the balloon from slipping during inflation. The balloon was inflated with normal saline using a balloon inflator.

Figure 1. (A) Chest radiograph showing short segmental smooth narrowing which involved both main bronchi. The trachea appears to be patent without focal lesion. Both lungs are clear. (B) Flow–volume graph of spirometry showing fixed upper airway obstruction pattern. (C) Chest scan with MinIP reconstruction technique showing segmental circumferential wall thickening involving bilateral proximal main bronchi, resulting in severe luminal narrowing.
at a dilating pressure of 5 bar for 15 sec, and then inflation repeated at a pressure of 8 and 10 bar. After the procedure, the bronchial lumens were significantly widened (Fig. 2B), the patient was eventually successfully taken off the ventilator and extubated. Oral prednisolone and cyclophosphamide were continued as maintenance immunosuppressive doses. Unfortunately, in late July 2015 the patient died from severe meningoencephalitis.

Discussion

The prevalence of GPA patients with airway stenosis have been reported to be 17.5%, and most of them have systemic manifestations simultaneously. The most prevalent stenotic area is the subglottis, whereas main bronchial stenosis is rarely seen. Bilateral main bronchial stenosis was reported in only 11.4% of airway stenosis cases [2,3].

Sixty percent of the patients with bronchial stenosis presented with no symptoms and were diagnosed by imaging or bronchoscopy performed for other reasons [3]. However, presentations in symptomatic patients may mimic other obstructive airway diseases. One of the interesting findings in this patient was the fixed obstruction pattern of flow–volume loop that was similar to a single obstruction in a more common presentation of GPA, subglottic stenosis. Computerized tomography scan and flexible bronchoscopy are required to identify the exact site of the lesions in patients who are showing this pattern of obstruction.

Acute respiratory failure from airway obstruction in GPA is uncommon. Most of the reported failures result from subglottic or tracheal obstruction [4]. Acute respiratory failure from bilateral main bronchial stenosis is rare.

Treatment of tracheobronchial GPA consists of medicamentous and non-medicamentous therapy. Pharmacological treatment of severe tracheobronchial disease should be a combination of oral glucocorticoids and cyclophosphamide [1]. Interventional bronchoscopy, including dilatational therapy (balloon, mechanical dilatation by rigid bronchoscope or Bougie dilator), laser ablation, electrocautery, argon plasma coagulation, cryotherapy, and stent placement, is beneficial in managing GPA tracheobronchial stenosis (TBS) [1,2]. These bronchoscopic procedures may be combined with other local adjuvant therapies such as intralesional injection of steroids or topical application of mitomycin-C [2]. The interventions could be performed using rigid or flexible bronchoscopy, depending on the availability and expertise of the physician. Bronchial balloon dilatation has been reported to be effective in treating benign TBS, including GPA [5]. Our report presents the benefits of BBD to rapidly relieve airflow limitation and allow the patient to be weaned from mechanical ventilation.
ventilation. However, recurrence of TBS is common and two thirds of the relapses occur during GPA remission [3]. Many patients require multiple interventions and silicone stents for long-term patency of the airway [1].

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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