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Case report

Biphasic flow-volume loop in granulomatosis with polyangiitis related unilateral bronchus obstruction

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ABSTRACT

Spirometry flow-volume measurement is used routinely in the outpatient setting to rule out obstructive lung diseases. Biphasic flow-volume loop is a classic presentation of unilateral bronchial stenosis due to multiple etiologies and it should raise clinical suspicion. Granulomatosis with polyangiitis (GPA) is a systemic inflammatory condition with pulmonary manifestations that may be infiltrative (e.g., pneumonia), hemorrhagic, and may rarely cause bronchial stenosis. Herein, we present a case of GPA-related, bronchial obstruction that caused biphasic flow-volume loop along with a literature review.

1. Introduction

Unilateral obstruction of the bronchus produces a classic biphasic flow-volume curve. In 1990, Gascoigne et al. first demonstrated a biphasic curve of flow-volume loop in 2 patients [1]. Unilateral obstruction of the bronchus can be caused by bronchomalacia, sarcoidosis, relapsing polychondritis, bronchial carcinoma, or Macleod's syndrome, amongst others (Table 1). A biphasic pattern present in patients, post-lung transplant, can indicate obstruction due to granulation tissue formation at the site of anastomosis [2]. Granulomatosis with polyangiitis (GPA) involving the tracheobronchial tree can also lead to unilateral bronchial stenosis. Herein, we present a case of a patient with GPA presenting with a biphasic flow-volume curve due to stenosis of the left main-stem bronchus, which improved upon bronchial dilatation.

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Table 1 – List of causes of unilateral bronchial obstruction causing biphasic flow-volume loop.

Developmental Cartilagenous Ridge [1]
Single Lung Transplantation [1]
Post Lung transplant – Due to development of granulation tissue at site of anastomosis [2]
Sarcoidosis [7]
Tracheobronchomalacia, Relapsing polychondritis [6,8]
Granulomatosis with polyangiitis [9]
Amyloidosis [1]
Bronchial carcinoma [5]
Unilateral lung emphysema (Macleod's syndrome) [5]

2. Case presentation

A 21-year-old male with a history of GPA was seen at our outpatient clinic complaining about shortness of breath. The patient had been diagnosed with GPA 2 years prior and he presented with arthritis, hematuria, and hemoptysis. He was found to be in acute renal failure with biopsy-proven, IgA-positive necrotizing and crescentic glomerulonephritis. Proteinase 3 antibody was found at the very high level of 141.8 AI (normal $\leq$ 1.0 AI). At that time, chest x-rays were unremarkable but computer tomography (CT) scans of the chest showed multifocal airspace opacities that were suggestive of alveolar hemorrhaging. He was treated with pulse dose steroids with a long steroid taper, induction with rituximab 375.0 g/m^2 4 $\times$ weekly, followed by methotrexate maintenance therapy. Over the course of 2 years, the patient had multiple recurrences of vasculitis with pulmonary infiltrates and received 2 more cycles of rituximab for GPA flares. At the time of this most recent presentation it had been 5 months since his last dose of rituximab; he was also taking prednisone 5.0 mg/day. The patient had stable vital signs and 99% oxygen saturation on room air. On lung auscultation, the patient was wheezing during forced exhalation. The remainder of the exam was unremarkable. Laboratory tests revealed blood urea nitrogen levels of 21.0 mg/dL, creatinine levels of 1.39 mg/dL, erythrocyte sedimentation rate (ESR) was 2 mm/hr, and c-ANCA and p-ANCA were both negative.

Pulmonary function test (PFT) revealed a severe obstructive disease with forced expiratory volume in 1 second (FEV1) of 1.95 L (35% of the predicted). The forced vital capacity (FVC) was 5.89 L (88% of predicted; FEV1/FVC of 0.33). The flow-volume loop showed a characteristic biphasic pattern (Fig. 1). Unilateral obstruction of the bronchus was suspected, given the resultant shape of the flow-volume loop. Chest CT scans revealed significant stenosis of the left mainstem bronchus (Fig. 2). Diagnosis of bronchial stenosis was also confirmed via bronchoscopy. Subsequently, the patient had undergone a bronchial balloon dilatation of the stenosis with the bronchus dilated from 4 to 11 mm at a different center. Biopsy was not preformed due to suboptimal biopsy conditions and risk of further granulation. A stent was not placed due to fear of propagation and further granulation. Patient pulmonary symptoms improved after the procedure. A follow-up CT scan revealed a dilated left mainstem bronchus. At an outpatient follow-up visit after 3 months, flow-volume measurement was performed. FEV1 increased to 4.33 L (79% of predicted FEV1) and FVC increased to 6.39 (95% of predicted FVC); FEV/FVC ratio was 0.68. Patient flow-volume loop showed significant improvement (Fig. 3); pre- and post-bronchial dilatation flow-volume loop measurements are shown in Table 2. The patient continued to follow-up with a rheumatologist for GPA treatment and continued to receive rituximab at 6 months intervals.

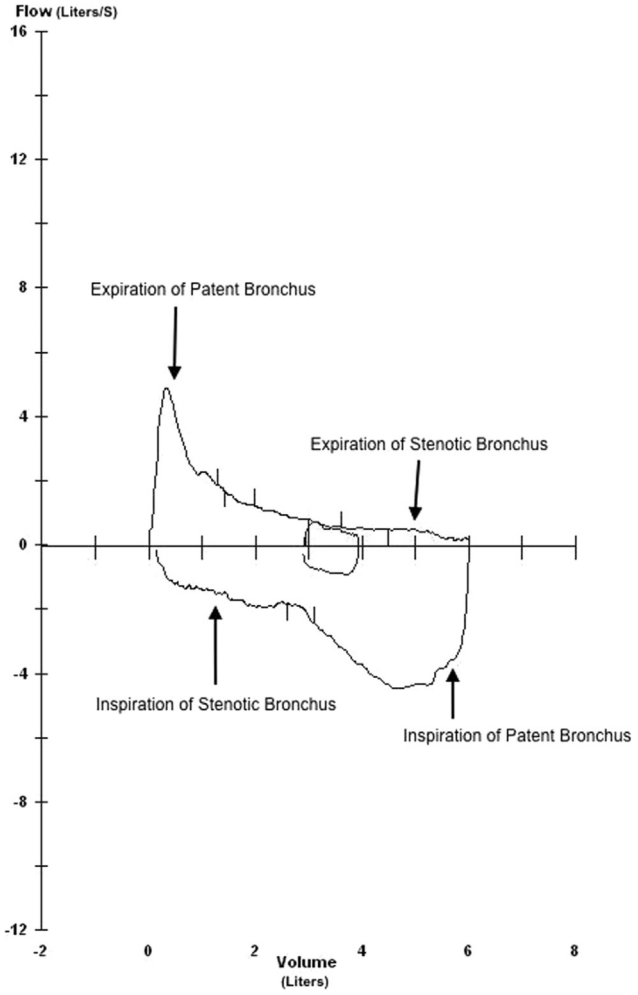

Fig. 1 – Pre-operative biphasic flow-volume curve.

3. Discussion

Spirometry with flow-volume loop curve is a routine outpatient procedure that evaluates patients with airway obstruction symptoms. Patients can present with dyspnea, wheezing, and coughing due to obstruction of the bronchus. The value of the shape of the flow-volume loop curve in maximal inspiration and expiration can help determine the level of obstruction and if the condition is fixed or dynamic [3]. Williams et al. studied the patterns of the flow-volume loop curve in patients with unilateral bronchial obstruction [4]. Patients with unilateral bronchial obstruction present with the classic biphasic flow-volume loop in both the expiratory and inspiratory phases; this is similar to how it presented in our patient.