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Case report - Thoracic non-oncologic

Bronchoplastic procedure for an unusual indication – Wegener’s granulomatosis

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Abstract

Wegener’s granulomatosis (WG) is a systemic vasculitic condition that commonly affects the lung and kidneys. With improvement in medical therapy, airway complications are increasingly encountered and are difficult to manage. Here, we present a case whereby a patient presenting with airway complication is successfully treated with surgery.

Keywords: Wegener’s granulomatosis; Bronchoplastic procedure; Thoracic surgeons

1. Case history

Wegener’s granulomatosis (WG) is a systemic vasculitic condition characterised by the presence of circulating anti-neutrophil cytoplasmic antibodies (ANCAs) affecting small- and medium-size blood vessels leading to end organ damage. The organs most commonly affected are the lungs and the kidneys. Treatment of this condition consists mainly of immunosuppression. Historically, thoracic surgeons play a role in diagnosing this condition through obtaining biopsy samples of the lung [1]. With the improvement in medical treatment, complications of WG involving the airways are increasingly seen. It is in this circumstance that thoracic surgeons have a new role in the treatment of this condition.

Here, we present a patient with stenosis of the right bronchus intermedius as a result of WG despite medical therapy, who was successfully treated with surgery. This case illustrates the novel role of surgery in managing this condition.

A 31-year-old man was diagnosed with WG in his early twenties. This patient was treated with a combination of immunosuppression consisting of steroid and mycophenolate mofetil. A referral to the tertiary thoracic centre was made when the patient started complaining of wheezing. The wheezing was mainly nocturnal and made worst on lying in the right lateral position. Bronchoscopy revealed severe stenosis of the right bronchus intermedius (Fig. 1). Computed tomography performed reveal several cavitations confined to the right middle and lower lobe with severe parenchymal destruction. As the patient had been on maximal medical therapy prior to referral, further escalation was thought to be unhelpful. After a multi-disciplinary discussion, it was decided that surgical approach may be the most appropriate.

This patient was brought to the operating theatre for resection of the stenotic bronchus intermedius segment and to assess the need for parenchymal resection of the diseased right middle and lower lobe. Intra-operatively, the lung parenchyma was found to be preserved. The right bronchus intermedius was then opened just distal to the right upper lobe bronchus and the stenosis identified. Next, the bronchus intermedius was opened proximal to the right middle lobe bronchus and the stenotic segment was found to be limited to the bronchus intermedius sparing the right middle lobe and right lower lobe bronchus. Hence, a bronchoplastic procedure consisting of resection of the right bronchus intermedius and end to end anastomosis of the right middle lobe, right lower lobe bronchus to the right upper lobe bronchus was carried out (Fig. 2). Anastomosis was constructed with 3/0 polypropylene suture using an interrupted suture technique. This patient’s post-operative recovery was uncomplicated and he was discharged 1 week after the procedure. The patient remains well on routine 6 weeks postoperative review.

2. Discussion

WG is a systemic vasculitic condition first described in 1936 by Dr Friedrich Wegener. It is characterised by granulomatous inflammation affecting the small- and medium-sized arteries and the presence of ANCAs. ANCAs activate neutrophils, increase neutrophil adherence to vascular endothelium, and lead to neutrophil degranulation causing damage to the vessel wall. The aetiology for the production of ANCAs is unknown, although some drugs have been implicated [2].
WG most commonly affects the upper respiratory tract, the lungs and the kidneys. Occasionally, it can also affect the skin, nervous system and the joints. In the lungs, WG leads to the development of cavitations and parenchymal destruction. Less commonly, the tracheo-bronchial tree is involved and become stenosed. Stenosis of the tracheo-bronchial tree is subtle and hence difficult to diagnose. On the other hand, when the diagnosis is made, the physician is faced with limited treatment options.

Thoracic surgeons traditionally play a role in WG by providing lung biopsy samples for diagnosis. However, recent reports suggest that thoracic surgeons are increasingly involved in providing therapeutic options in WG. This includes bronchoscopic dilation and stenting of stenotic airway and intralesional steroid injection [3, 4]. With the increasing popularity of bronchoplastic surgical techniques, the role of thoracic surgery in the treatment of WG may expand especially in dealing with stenosis of the tracheo-bronchial tree. In this report, we illustrate a case where stenosis of the tracheo-bronchial tree secondary to WG despite optimal medical treatment is successfully treated surgically using complex bronchoplastic technique. With the advancement in technical ability of thoracic surgery, more complex bronchoplastic procedure may be undertaken in the future to improve the management of stenotic airway seen in WG. Hence, we advocate that surgeons involved in managing this condition should be familiar with bronchoplastic techniques in order to optimally manage the condition.

References

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