Case Report

Successful Stenting in Endobronchial Wegener’s Granulomatosis

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INTRODUCTION

Wegener’s Granulomatosis (WG) is a multisystem disorder characterized by granulomatous necrotizing vasculitis. Classic Wegener’s granulomatosis is a triad of necrotizing angitis of the upper and lower respiratory tract and focal glomerulonephritis of the kidney. The classic respiratory feature is multiple pulmonary nodules on chest radiograph. In many cases extensive medical evaluation and laboratory test have proven non-diagnostic. In 1966, Carrington and Liebow introduced the concept of “limited Wegener’s” granulomatosis to identify otherwise classic vasculitis lacking renal involvement. Limited Wegener’s granulomatosis has a better prognosis than classic disease but it may be extremely challenging to recognize and diagnose.

We report an unusual case of limited Wegener’s granulomatosis presenting with focal endobronchial WG with lobar collapse requiring stenting.

CASE REPORT A 19 year old female student presented to Otorhinolaryngology in June 1997 with a 3-week history of nasal obstruction, anosmia, headache, post-nasal drip and cough, unresponsive to recurrent antibiotic courses. X-Ray of paranasal sinuses revealed both maxillary sinus opacity. She was admitted in January 1998 for bilateral antral washouts and nasal endoscopy. Postoperatively, she developed fever, malaise, anorexia and unexplained weight loss. CT scan of paranasal sinuses revealed pansinusitis. She had bilateral functional endoscopic sinus surgery without much benefit. A CT Scan of brain excluded intracranial abscess. Revision endoscopic sinus surgery, performed 9 days later, revealed pus with necrotic material in the maxillary sinuses. Despite repeated sinus drainage procedures and intravenous broad-spectrum antibiotics during her hospitalisation, she continued to be febrile with weight loss. She had persistently elevated C – reactive protein [130 – 393 mg/l]. Her Westergren erythrocyte sedimentation rate was 110mm/hour. All cultures were negative. No granuloma or fungus was observed on biopsies. Initial autoimmune and vasculitic tests demonstrated no elevation in autoantibodies. She developed a normocytic anaemia, transient polyarthralgia and destructive inflammation of her nasal bridge.

Despite the initial absence of granuloma on histology or Anti Neutrophil Cytoplasmic Antibody (ANCA) in serum, a provisional clinical diagnosis of Wegener’s granulomatosis was made. The patient was commenced on high dose oral steroids and co-trimoxazole. Steroid therapy resulted in prompt response and rapid clinical improvement, evident within 24 hours.

Indirect serum immunofluoresence in early February 1998 showed an atypical positive pattern for cANCA and Antiproteinase 3 level 6.2U/L [Normal <2]. Cyclophosphamide was added to her management regime. Rapid symptomatic improvement followed and she was discharged home.

In June 1998 she presented acutely unwell with shortness of breath and fever. A chest radiograph revealed complete collapse of the left lower lobe (fig.1). Bronchoscopy confirmed

![Fig 1. Chest X-ray pre-stent.](image-url)
complete occlusion of the left main bronchus. Treatment was commenced with intravenous methylprednisolone, cyclophosphamide and antibiotics. Repeat bronchoscopy and biopsy of firm tissue at the stenosed left main bronchus showed superficial fragments of oedematous and reactive mucosa with extensive squamous metaplasia. There was no dysplasia, malignancy, granulation, vasculitis or fungus. Rigid bronchoscopy with laser ablation of the stenotic segment followed by dilatation was performed. Rapid improvement ensued.

Her health deteriorated within three weeks, with restenosis of the left main bronchus. Repeat bronchoscopy and laser ablation of stenotic segment followed by dilatation gave immediate relief. Restenosis occurred and the cycle continued. In a 2-month period, a total of 9 dilatations were carried out, several with laser resection. Each gave transient symptomatic relief.

The patient proceeded to stenting in October 1998. Rigid bronchoscopy provided accurate measurement of the extent of the stenotic segment of left main bronchus. An endobronchial stent was deployed after laser ablation and balloon dilatation (fig. 2).

She improved dramatically following stenting. Follow up at the respiratory outpatient clinic enabled slow tapering of her steroid therapy to zero by July 2000. All bloods were normal and azathioprine therapy was discontinued in October 2000. She underwent nasal reconstructive surgery in 2001. She was able to complete her studies and was married early in 2003. She underwent nasal reconstructive surgery in 2001. She was able to complete her studies and was married early in 2003. Her only complaints during this 5-year period were occasional upper respiratory tract infections, which responded well to short courses of oral antibiotics.

A deterioration in FEV1 late in 2003 raised suspicion of restenosis (FEV1 3.4L May 2002 - FEV1 1.75L November 2003). Bronchoscopy in November 2003 demonstrated a well epithelialised, stented left main bronchus with mobile, occluding but not actively inflamed, tissue at the distal end of stent. In the coming weeks she was monitored closely. At surgical review in February 2004 she was 8 weeks pregnant and subjectively well. Intervention was postponed.

A further acute presentation with a left lower lobe pneumonia occurred in April 2004. Subsequent bronchoscopy revealed a circumferential stenosis of greater than 50% at the origin of the left main bronchus. Histology confirmed recurrence of acutely inflamed granulation tissue and laser ablation was undertaken in May 2004.

She had a baby girl by caesarean section in September 2004. In 2005 laser debulking of the endobronchial lesion has been performed twice. A repeat biopsy in January 2005 revealed further stent hyperplasia with granulation tissue. Both treatments have provided symptomatic relief.

**DISCUSSION**

In this patient the upper airway disease due to Wegener’s Granulomatosis responded to corticosteroids, cyclophosphamide and co-trimoxazole. Endobronchial WG is an uncommon manifestation of the disease. Koyama et al (2003) reported the incidence of endobronchial involvement to be as low as 16%. Hirsch et al (1992) further stressed the rarity of endobronchial WG. They described a patient in whom the only manifestation of WG is severe proximal bronchial stenosis, which developed despite management with oral steroid and cyclophosphamide. That case responded well to IV cyclophosphamide and oral co-trimoxazole whereas our case required further intervention. A further case report by Breton et al (1986), illustrated the difficulty in diagnosing WG from such a presentation and emphasise the need for tissue biopsy. These highlight the importance of awareness of this infrequent but potentially life-threatening feature of the condition.

Subsequent to presentation with left main bronchus occlusion, our patient underwent repeated dilatation and laser ablation therapy. Each of these afforded her only temporary relief of airway obstruction. Several authors suggest that balloon dilatation is safe, efficacious and cost-effective. It is easily performed and has few complications. Eagleton et al found repeated endoscopic dilatation effective for 18 months in a patient with endobronchial WG. Until now, no long-term follow up has been published. When combined with laser therapy, endoscopic dilatation may be acceptable as a first line measure to restructure the occluded airway but to further enlarge the airway and maintain patency, stenting is required.

Endobronchial stenting is not without complication. Problems include displacement of the stent and obstruction with secretions or granulation tissue (as in our patient). Less commonly the stent may perforate the airway wall, sometimes into the accompanying blood vessel. Most studies of endobronchial stenting demonstrate that they are an effective treatment modality for airway stenosis. The long term efficacy of endobronchial stents is superior to laser ablation, debulking or dilatation therapy though the long term patency of stents is uncertain.

Stents have been effective in achieving immediate resolution of respiratory symptoms from various tracheobronchial obstructions. In the majority of cases, however, the success
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of such a management option cannot be judged on such short term outcomes. Currently, the ideal course of action is unclear, as none are free from complications, nor able to consistently provide life-long patency.15

This patient’s endobronchial stenosis was successfully managed with endobronchial stenting which maintained airway patency for 5 years. No other reports were found with such encouraging longterm outcomes.

The authors have no conflict of interest

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