Bronchial Stenosis and Extensive Bronchiectasis due to Wegener’s Granulomatosis

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Dear Sir,

Much has been learned about pathogenesis and natural history of Wegener’s granulomatosis (WG). Likewise, our armamentarium of therapeutic options has increased considerably. Tracheobronchial involvement, however, remains a diagnostic and therapeutic challenge. Here, we briefly describe the case of a patient with biopsy-proven WG of the nose, paranasal sinuses and kidney who, while on treatment with cyclophosphamide and steroids, developed a stenosis of the right upper lobe bronchus and severe bronchiectasis necessitating surgical removal of the affected lobe.

A then 50-year-old male patient presented first in January 1997 with pneumonia, night sweats, and weight loss. On examination there was mild left hemiparesis. A computed tomography scan of the brain disclosed a 2-cm lesion of the internal capsule. Severe pansinusitis was also detected. Eventually, surgery of the paranasal sinuses was performed and histology of the specimen revealed granulomatous vasculitis. Prompted by nephritic urinary sediment and normal serum creatinine, urgent renal biopsy was performed revealing pauci-immune crescentic glomerulonephritis. Infiltrations of the right upper lobe, previously interpreted as pneumonia, failed to clear despite adequate antimicrobial chemotherapy. However, further workup of the thoracic abnormalities was deferred because the patient wished so. Augmented by positive assays for cytoplasmic antineutrophilic antibodies and anti-proteinase-3 antibodies, a diagnosis of WG with involvement of nose, sinuses, kidney and probably lung was made. Treatment with steroids and oral cyclophosphamide was initiated. Six months after the initial presentation, the patient presented again with recurrent pneumonia and hemoptysis. Chest x-ray showed linear infiltrations of the right upper lobe (fig. 1). A computed tomography scan showed extensive bronchiectasis of the right upper lobe (fig. 2). Vital capacity
and forced expiratory volume of 1 s (FEV₁) were 90 and 80% of the age-adjusted normal value, respectively. Bronchoscopy revealed stenosis of the right upper lobe bronchus; histology confirmed granulomatous inflammation in keeping with WG. Cultures of bronchoalveolar lavage remained sterile. Resection of the right upper lobe was performed after a trial of antimicrobial chemotherapy. The patient made an uneventful recovery and is currently well and in remission after he completed a 1-year trial of oral cyclophosphamide.

Pulmonary involvement of various types is common in WG. It may occur in the context of generalized vasculitis but disease activity may also be confined to the respiratory tract (‘limited WG’). Occasionally, bronchial WG may be the initial manifestation [1]. Among 77 patients with pulmonary WG reported by Cordier et al. [2], bronchial involvement was encountered in 30 patients but only 13 (16%) had bronchial stenosis disclosed by endoscopy. Daum et al. [3], in their 1995 single-center experience with tracheobronchial involvement in WG, reported bronchial stenosis in 13% of patients but only 2 patients had severe stenosis.

A broad variety of inflammatory [4] and infectious [5, 6] lesions may cause bronchial stenosis and thus mimic WG. Proving the diagnosis may be difficult since the yield of endoscopic biopsy may be unsatisfactory. Fortunately, tracheobronchial involvement in WG often responds to immunosuppressive treatment, sometimes surprisingly well [7]. Rarely, bronchial stenosis becomes the predominant clinical problem despite adequate treatment with good control of systemic disease activity. Anecdotal evidence suggests that endobronchial intervention with stent-graft placement is feasible and efficient in these cases [3, 8, 9]. Very rarely, however, bronchiectasis develops despite therapeutic efforts. We report such a case of bronchial stenosis due to WG with subsequent extensive bronchiectasis, a rare manifestation of WG successfully managed by surgical resection.

References

Fig. 2. CT scan demonstrating severe bronchiectasis of the entire right upper lobe.