Subglottic stenosis in granulomatosis with polyangiitis (Wegener granulomatosis)

A 36 year old woman presented with a 1 year history of nasal obstruction, exertional dyspnea with biphasic stridor and 10 kg unintentional weight loss. Laboratory investigations revealed that her hemoglobin level was 8.1 g/dL and erythrocyte sedimentation rate was 41 mmph. Her anti-neutrophil cytoplasmic antibodies were positive. Chest radiograph revealed a small faint patch over left upper lung. Sinoscopy revealed a roomy nasal cavity caused by atrophy of the mucosa, thick discharge, lots of dark yellow crusts, epistaxis and a large perforation of the nasal septum. These findings suggested atrophic rhinitis complicated with perforation of the nasal septum. These findings suggested atrophic rhinitis complicated with perforation of the nasal septum (FIGURE 1 arrows: perforation of the nasal septum, asterisk: atrophy of the inferior turbinate). Bronchoscopy revealed approximately 80% stenosis of the subglottis (grade III, Cotton-Myer grading system) (FIGURE 2 arrows) and long-segmental subglottic granulomatous masses involving the glottis (stage IV, McCaffrey classification system) (FIGURE 3 arrow). An impending life-threatening subglottic stenosis was noted. Therefore, the patient underwent ventilation bronchoscopy and laryngotracheal reconstruction with a T-tube (Montgomery®, Standard Safe-T-Tube™, size 11) stent below the vocal cord (Figure 4 arrow). Postoperatively, granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis, was diagnosed on the basis of clinical features and histological evidence of necrotizing vasculitis and granulomatous inflammation [1]. The patient’s dyspnea was greatly improved by the surgical intervention alone. The patient did not have any vasculitic manifestation except for respiratory tract at 1 year close follow-up. Guardiani et al. observed 31% patients with GPA presented with subglottic stenosis as part of their initial manifestation. The majority of patients had concurrent sinonasal involvement [2]. Jordan et al. observed 78% patients with GPA presenting with subglottic stenosis were women [3]. Subglottic stenosis is a common and significant presenting manifestation of GPA. Awareness of subglottic stenosis in patients with GPA is essential to avoid delayed diagnosis or life-threatening crisis.
REFERENCES

1. Lutalo PM, D’Cruz DP. Diagnosis and classification of granulomatosis with polyangiitis (aka Wegener’s granulomatosis). J. Autoimmun. 48, 94-98 (2014).
