Large airway involvement in relapsing polychondritis

Michael Greenstone, Elaine Baguley

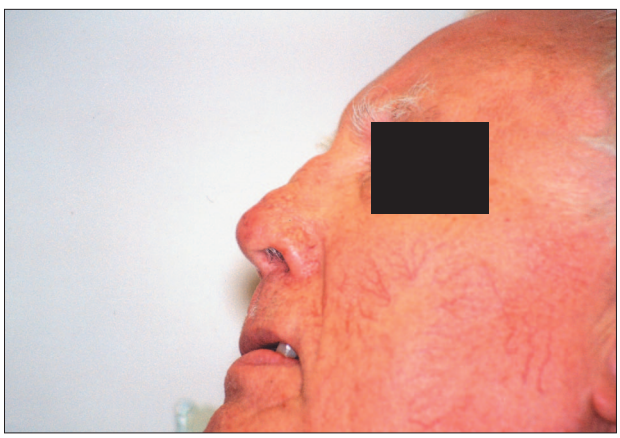


Figure 1. Collapse of the cartilaginous nasal septum with preservation of the proximal bony septum.

Figure 2. Inflammation of the ear with characteristic sparing of the inferior soft lobule.

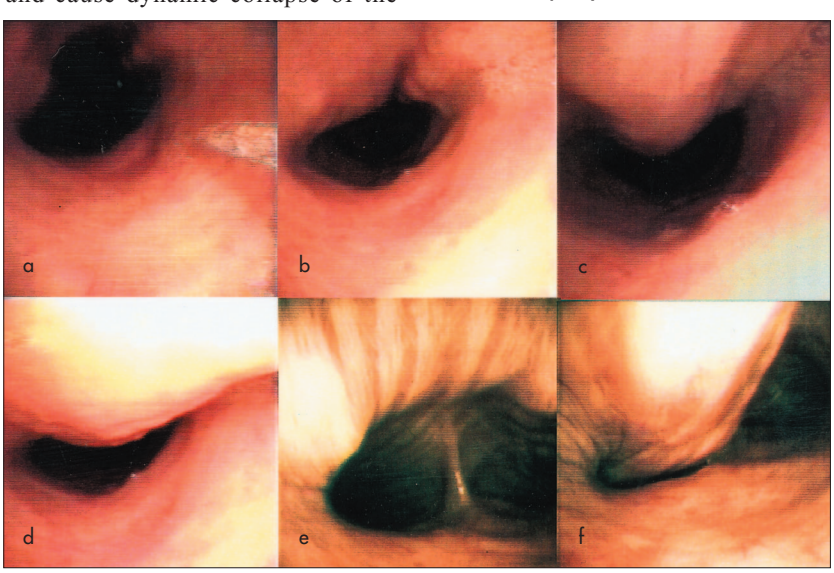
A 67-year-old man presented with conjunctivitis and deafness followed by bilateral uveitis, weight loss and a vasculitic rash. His nasal bridge had collapsed (*Figure 1*) and the upper part of his left ear was thickened and distorted (*Figure 2*).

Investigation showed anaemia with elevated inflammatory markers but negative antineutrophil cytoplasmic antibodies. Relapsing polychondritis was diagnosed and he was treated with corticosteroids and pulsed cyclophosphamide with improvement.

Eighteen months after presentation he developed increasing dyspnoea, stridor and evidence of an obstructive ventilatory defect. Bronchoscopy showed expiratory collapse of the posterior tracheal wall through a cartilaginous defect (*Figure 3a–d*) and occlusion of the right main bronchus at the level of the main carina (*Figure 3e–f*). During inspiration the appearances were normal. Symptoms improved after insertion of an endotracheal stent.

Relapsing polychondritis is a rare multisystem disorder characterized by recurrent episodes of inflammation of cartilage and proteoglycan-rich structures. Respiratory tract involvement is common and usually affects the nose, larynx and upper trachea. Inflammatory oedema can create airway narrowing and then destroy laryngeal, tracheal and bronchial cartilage and cause dynamic collapse of the large proximal airways. Involvement of the lower respiratory tract is one of the most serious complications of relapsing polychondritis; it carries a poor prognosis and may account for up to 50% of deaths. HM

Figure 3. Dynamic collapse of the trachea and right main bronchus: on expiration the posterior tracheal wall protrudes through the defect and narrows the lumen of the trachea to 30% of its normal diameter. The right main bronchus appears to occlude completely.



Dr Michael Greenstone is Consultant Physician in Respiratory Medicine and **Dr Elaine Baguley** is Consultant Rheumatologist, Hull Royal Infirmary, Hull HU3 2JZ

Correspondence to: Dr M Greenstone