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Pulmonary puzzle

ANSWER

From the question on page 483

Scar tissue has caused cicatricial supraglottic stenosis from histologically confirmed mucous membrane pemphigoid.

Dysphonia (from anterior glottic scarring) and dyspnoea (from posterior scarring) may progress to severe stenosis, stridor and life-threatening airway obstruction. Laryngeal stenosis predominantly affects children, either as a result of a congenital glottic web or papillomatosis. The subglottis is the most common site for a stricture secondary to an intubation injury. Inflammatory causes include croup, epiglottitis and laryngo-tracheobronchitis. Adult causes are shown in box 1.¹

Mucous membrane pemphigoid is a rare chronic systemic autoimmune disease affecting the mucous membranes. Antibodies bind to the basement membrane causing subepidermal bullae which rupture with scarring. There is a preponderance of females and the sixth decade.² Complications include ocular lesions (75% of patients) with blindness in up to 20%,³ oesophageal strictures, laryngeal stenoses (8%)⁴ and bronchial stenoses. Flow-volume loops may show airflow disruption. Ocular, laryngeal, oesophageal, nasopharyngeal and genital involvement are deemed high risk for progression carrying a worse prognosis,³ the anti-epigrilin (laminin 5) variant particularly being associated with malignancy.

Trials of management are few and inconclusive,⁵ and relapse and progression are common. Supraglottic stenosis may require repeated laser therapy, adjuvant mitomycin C or, ultimately, tracheostomy. In this case, the stenosis remains under review.

Supraglottic obstruction is illustrated as a rare cause of exertional dyspnoea.

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Box 1 Laryngeal obstruction or stenosis: aetiology and reported anatomical sites

Supraglottic

- ▶ Carcinomas (95% squamous)
- ▶ Relapsing polychondritis
- ▶ Mucous membrane pemphigoid
- ▶ Diphtheria
- ▶ Epiglottitis

Transglottic

- ▶ Carcinomas
- ▶ Papillomatosis
- ▶ Sarcoidosis
- ▶ Tuberculosis
- ▶ Perichondritis and radionecrosis
- ▶ Fracture

Subglottic

- ▶ Carcinomas
- ▶ Wegener granulomatosis
- ▶ Intubation injury

Other

- ▶ Carcinoid, amyloid, Kaposi sarcoma, metastases, gastro-oesophageal reflux.

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