UPPER AIRWAYS INVOLVEMENT IN WEGENER’S GRANULOMATOSIS. A CASE REPORT AND REVIEW
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Abstract:
Upper airway involvement is an important and often missed complication of Wegener’s granulomatosis. A 29 years old girl with known Wegener’s granulomatosis presented with progressive dyspnoea, brassy cough, hoarse voice and mild stridor. Chest was normal on auscultation. Chest radiograph was normal but pulmonary function tests showed extra-thoracic compression and CT scan confirmed inflamed and narrow glottis. She was treated with high dose steroids and intravenous pulses of cyclophosphamide. At follow-up, symptoms, repeat pulmonary function test and CT scan all showed improvement.

Key words: Wegener’s granulomatosis, laryngeal involvement, stridor

Introduction:
Wegener’s granulomatosis (WG) is a necrotising vasculitis, which often involves the sinuses, pulmonary parenchyma and kidneys (1). Upper airway involvement is an important and often missed complication of WG. We present a young lady with established WG who had upper airway involvement and review the relevant literature.

Case Report:
A 29 years old lady who has been under our care for Wegener’s granulomatosis since 1990. She was initially treated with steroids and cyclophosphamide. Due to persistent amenorrhea, cyclophosphamide was stopped and methotrexate initiated. In 2003 she presented to the rheumatology clinic with persistent cough and dyspnoea.

On examination, she had mild dyspnoea at rest, stridor and brassy cough. There was nasal crusting and collapsed nasal bridge (Figure 1). Chest was otherwise normal. Other systems were normal.

![Figure 1: Plain CT scan through the larynx shows soft tissue fullness along the vocal cords with mild narrowing of the laryngeal lumen.](image)

Investigations done were: 13 gm/dl, WBC: 12100/cumm, ESR – 32mm/hr, Creatinine – 0.8 mg/dl and urinalysis – Normal. Chest radiograph was normal. Pulmonary function tests were suggestive of extra thoracic upper airway obstruction (figure 2). Inspiratory and expiratory loops where flattened, Empey’s Index was 11.69 and FEF 50/FIF 50 was 1.27 CT scan chest with virtual bronchoscopy
showed narrowed glottis during inspiration and expiration (figure 3). Bronchoscopy showed sluggishly moving vocal chords, swollen and edematous subglottic space, which was circumferentially narrowed and could not be entered.

She was treated with 3 doses of pulse IV methylprednisolone (1g each), followed by oral steroids (1 mg/kg taper) and IV cyclophosphamide pulses. Subsequently she was continued on oral cyclophosphamide.

At 6 months follow-up she has improved.

The stridor and brassy cough were less. The pulmonary function tests had improved as follows: FVC improved from 74.9 to 78.3% and PEFR improved from 51% to 78%. The flow volume showed no upper airway obstruction, Empey’s Index improved to 8.9 and FEF 50/FIF 50 was 0.83. She is now on 10 mg of prednisolone and 150 mg oral cyclophosphamide.

Discussion:

Wegener’s granulomatosis (WG) is granulomatous vasculitis of the small and at times medium sized vessels. Involvement of ear, nose, throat region, pulmonary parenchyma and kidney are all well described.

Laryngotracheal involvement, which occurs in up to 16% of patients, may be missed. It may be asymptomatic but its manifestations may range from subtle hoarseness to life threatening stridor and upper airway obstruction (2). The characteristic lesion is subglottic stenosis. Chest radiography may be normal.

Pulmonary function tests with flow volume studies show flattened inspiratory loop in variable
extrathoracic obstruction and flattened inspiratory and expiratory loops in fixed obstruction. CT scan with or without virtual bronchoscopy could define the lesions. Direct laryngoscopy is often essential and reveals acute erythematous friable mucosa or scarring. Circumferential narrowing below the cords may be seen. Bronchoscopy confirms the above findings.

In the early stages steroid and immunosuppressive agents like cyclophosphamide are used, but only 20% of patients respond whilst 80% have fixed obstruction. Bronchoscopic intralesional injections of steroids may be of some benefit (6). Laser (CO₂) excision of the subglottic stenosis has been tried (6). Airway dilatation has been tried with silicone or metal stent placements but prognosis remains guarded (6). Definitive therapy for resistant fixed obstruction could entail open airway reconstructive procedures such as laryngotracheoplasty and cricotracheal resection with primary anastomosis (6, 7).

References: