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Airway Management in case of Granulomatosis with Polyangiitis

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Clinical History

A 33-year-old female, complains of dyspnea for 1 month, insidious in onset, progressive, initially MMRC 0 to MMRC 4 (dyspnea on rest), (Modified Medical Research Council Grading), no postural variations, no aggravating and relieving factors.

She also complains of cough with expectoration, insidious onset, progressive, moderate quantity sputum, yellow colored sputum, no postural variations, no diurnal variations.

No h/o fever, ear, nose throat problems.

No h/o chest pain, palpitations, fatigue, sweating, pedal edema.

No h/o abdominal pain, vomiting, diarrhea.

Past History:

K/c/o AAV (ANCA Associated Vasculitis) for 5 years on regular treatment.

K/c/o HTN for 5 years on T. LOSARTAN 50mg.

Not a k/c/o DM, Epilepsy, Asthma, TB, Thyroid derangement

Examination and Investigations

Examination:

GPE:
Middle aged, moderately built and nourished, well oriented to time, place and person, alert, conscious and cooperative.

No Pallor, Icterus, Cyanosis, Clubbing, Lymphadenopathy, Edema.

**Vitals:**

**Pulse Rate:** 78 bpm,

**BP:** 110/70 mmHg,

**RR:** 17 cpm,

**Temperature:** 97 *F

**Systemic Examination:**

**CVS:** S1, S2 heard, no murmurs.

**RS:** B/L A/E+,

**B/L NVBS +**

Right Infrascapular area crepts,

**rhonchi ++**

**PA:** Soft, non-tender, no organomegaly

**CNS:** NFND

**Eye:** Ciliary Staphyloma ++ in both eyes (Scleral thinning in Left eye from 9 to 5 o clock,
Corneal thinning from 5-6 o clock, Scleral perforation treated with graft 2 years back),

Diminished vision in Right eye not improving on pin hole, Pupils round reactive in both eyes.

**Fundus:** Normal
**Investigations:**

**TLC:** 14300 cells/ cu mm

**PT:** 11.8s,

**aPTT:** 27.6s, INR:0.79

**cANCA:** POSITIVE

**pANCA:** Negative

**ANA Profile:** Negative

**Laryngoscopic Evaluation:** Sub glottic stenosis found with obstruction more than 75% of lumen of airway

**Diagnostic Nasal Endoscopy:** Deviated nasal septum with nasal spur to left nasal cavity.

**HRCT Thorax:**

Loss of volume in B/L upper lobes with areas of fibrosis. Right apical pleural thickening noted.

2 nodular lesions in posterior basal segment of Right lower lobe and another nodular lesion in left apico-posterior segment.

Areas of consolidation in superior and posterior basal segments of Right Upper Lobe and medial segment of Right Middle lobe.
Irregular thick-walled cavities in right upper lobe, posterior and lateral basal segments of right lower lobe and apico-posterior segment in left upper lobe.

**ECG**: Normal

**2D ECHO**: Normal

![X Ray Neck lateral view](image)

Sub-glottic stenosis seen in laryngoscopic examination

**Final Diagnosis**

Granulomatosis with Polyangiitis and Pulmonary involvement

Subglottic stenosis

Systemic Hypertension

**Discussion**
Key Points:

Granulomatosis with Polyangiitis (GPA), formerly known as Wegener’s Granulomatosis, is an autoimmune disorder that affects small blood vessels. (1) It is included in ANCA Associated Vasculitis along with Churg-Strauss Vasculitis and Microscopic Polyangiitis.

Symptoms include:

- Chronic Sinusitis
- Breathlessness
- Hemoptysis
- Blood in the urine.

The major autoantigens to which ANCA are directed within neutrophils and monocytes include two enzyme proteins identified in the 1980s as autoantigens, namely,

- Proteinase-3 (PR3)
- Myeloperoxidase (MPO) (2)

Tracheobronchial Stenosis are serious complications of GPA which can arise at any stage of the disease. Timely diagnosis plus optimal treatment and follow-up remain unmet needs. (3)

T Tube has to be placed in patients of GPA if subglottic stenosis has set in as it can compromise airway. It will lead to complete cut off of ventilation and is fatal.

Ideal treatment is LASER EXCISION but since the patient presented with acute stridor and in view of limited resources, tracheostomy with T Tube insertion was done as a life saving measure.

Acknowledgements: none
References:

1) API Textbook of Medicine

2) Kelley Textbook of Rheumatology

3) Frequency, Treatment, Evolution, and Factors Associated with the Presence of Tracheobronchial Stenoses in Granulomatosis with Polyangiitis. Retrospective Analysis of a Case Series From a Single Respiratory Referral Center

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