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Case Report

Wegener's Granulomatosis and Subglottic Stenosis - A Case Report

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Abstract

Wegener's Granulomatosis also known more recently as Granulomatosis with polyangiitis, is a systemic autoimmune vascular disorder which usually affects small and medium sized vessels of the body. Often there is presence of necrotising granulomatosis resulting in formation of localised granulomas along with the tissue damage [1,2]. It commonly involves various organ systems like Nose, Lungs, Nasal sinuses, Trachea, Kidneys, Heart, Skin, Eyes, Ear and various body joints often resulting in cough, chest pain, haemoptysis, sinusitis, otitis, rhinitis, subglottic stenosis, nasal septum perforation, nasal mucosa ulcers, glomerulonephritis, pericarditis, skin lesions [3].

Here we report a case of Wegener's Granulomatosis scheduled for Bilateral Functional Endoscopic Sinus Surgery (FESS) and Bilateral Myringotomy with Grommet Insertion requiring General Anaesthesia.

Keywords: Wegener's Granulomatosis; Bilateral Functional Endoscopic Sinus Surgery (FESS); Bilateral Myringotomy

Introduction

Wegener's Granulomatosis also known more recently as Granulomatosis with polyangiitis, is a systemic autoimmune vascular disorder which usually affects small and medium sized vessels of the body.

Case Report

A 36 year old female, diagnosed with Wegener's granulomatosis since 5 years, came with chief complaints of recurrent headache and nasal congestion on and off for 4 years, bilateral hearing loss slowly progressive over past 4 years. She had visited various ENT specialists for her complaints and was started on Tab Prednisolone 2.5 mg 4 years ago gradually increased to 10 mg OD at present and Tab Cyclophosphamide 50 mg alternate days for past 6 months. Audiometry showed bilateral conductive hearing loss. The patient was posted for Bilateral FESS and myringotomy with grommet insertion for symptomatic relief. She didn't give any history of breathlessness, chest pain, breathlessness on exertion, haemoptysis, any skin lesions or joint pain.

On general examination patient was comfortable. Vitals were stable. On systemic examination chest was clear, bilateral air entry present, no adventitious sounds were heard. Heart sounds were normal. Bilateral decreased hearing was present. Airway examination showed Modified Mallampati grade 1, mouth opening approximately 7 cms without any restriction of neck movements. Audiometry showed bilateral conductive hearing loss. Complete hemogram, Renal function tests, Liver function tests, Coagulation profile, Urine routine microscopy, Serum cortisol levels, Electrocardiogram, 2D echo were all normal. Chest X-Ray PA view did not reveal a narrowed airway shadow (Figure 1). Her ESR levels were mildly raised and Antinuclear antibody (ANA) test and Antineutrophil cytoplasmic autoantibody (c-ANCA) test were positive. HRCT Paranasal sinuses showed chronicity with soft tissue plugging of bilateral maxillary ostium and progression of soft tissue plugging of right spheno ethmoidal recess and left side ethmoid sinusitis. A previous HRCT chest didn't reveal any airway abnormality.

Patient and family were explained in detail about anaesthesia, surgery procedure and associated expected complications. Informed and valid consent was obtained. On the morning of surgery anaesthesia machine and circuit were thoroughly checked, anaesthesia drugs were loaded and kept ready, emergency drugs

kept standby, difficult airway cart was kept ready in view of anticipated difficult airway. A wide bore IV line with 18G was secured. Monitors-ECG, BP, Saturation probe, end tidal CO_2 were attached, and patient was preoxygenated for at least 3 minutes with 100 percent oxygen. Inj. Glycopyrrolate 0.005 mg/kg body weight, Inj. Fentanyl 2 mcg/kg body weight, Inj. Ondansetron 15 mg/kg body weight and Inj. Hydrocortisone 100 mg for steroid coverage IV given. Patient induced with Inj. propofol 2 mg/kg body weight IV with titrated doses and mask ventilation ensured. After confirming mask ventilation without any resistance Inj. Succinyl choline 2 mg/kg IV given. Direct laryngoscopy done with Macintosh Laryngoscope with visualisation of cormack lehane grade 1, intubation was attempted with cuffed portex size 6.5 mm endotracheal tube but was unsuccessful, smaller sized tubes also could not be passed through below the vocal cords. Patient was mask ventilated till effect of succinyl choline wore off and awake fibre optic intubation was attempted which showed severe (grade 3) subglottic stenosis (Figure 2). Surgery was postponed till further evaluation.

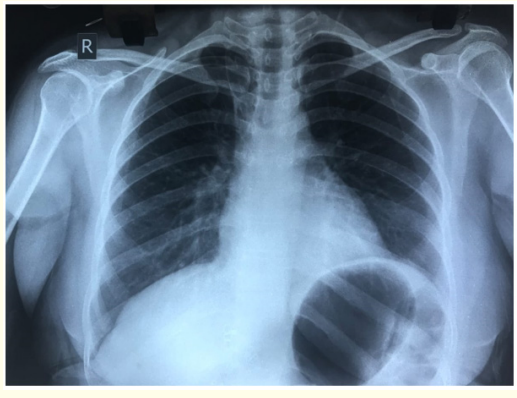

Figure 1

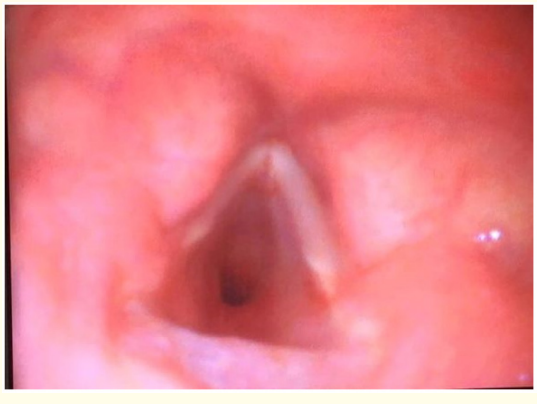

Figure 2

Patient and her family were counselled for CO_2 laser therapy for relieving subglottic stenosis at a later date.

Discussion

Wegener's Granulomatosis is a multisystem autoimmune disorder characterised by granuloma formation, vasculitis of both arteries and veins and glomerulonephritis [4]. It is now a recognised disease in Indian population. Earlier many cases were masqueraded to be as pulmonary tuberculosis before the advent of ANCA tests. Patient may present with a large spectrum of symptoms like upper airway involvement in the form of rhinitis, sinusitis, epistaxis, saddle nose deformity, nasal septum perforation, lower airway involvement like subglottic stenosis, cavitary lesions in lungs, haemoptysis, shortness of breath, interstitial infiltrates, otologic symptoms like otitis, tinnitus, eustachian tube dysfunction, eye involvement in the form of uveitis, scleritis, conjunctivitis, keratitis, kidney involvement in the form of glomerulonephritis, joint involvement etc [5]. Anaesthesia management in such patients depends upon the extent of various organ systems involved. These patients are usually potent candidates for difficult and reactive airway. Due to the presence of granuloma and inflammation, there are high chances of airway obstruction, oedema and bleeding. Thus, for giving General anaesthesia to such a patient, a thorough and meticulous examination of the airway including an Indirect laryngoscopy is a must. X-ray Neck AP and Lateral view, Chest x-ray AP and Lateral view and HRCT chest should be done to look for any obstructions or lesions in the upper and lower airway. If there are any complaints of respiratory compromise a preop Arterial Blood Gas analysis should be done. Patient and relatives should be thoroughly counselled and an informed consent for post op mechanical ventilation/ tracheostomy should always be obtained for emergency scenario. Inside operation theatre difficult airway cart consisting of appropriate sized facial masks, Oral airway, Nasopharyngeal airway, Endotracheal tube different sizes cuffed and uncuffed, Micro laryngeal tubes, different size Macintosh blades with 2 working scopes, stylet, Bougie, video laryngoscope with D blade, fibreoptic bronchoscope should always be kept ready. Instruments required for cricothyroidotomy or tracheostomy should be readily available. ENT surgeon should be present inside the OT at all times. Even in patients posted under regional anaesthesia difficult airway cart should be kept ready [6].

Supraglottic airways like laryngeal mask airways can also be used in surgeries where it is safe to use them without any danger of further obstruction of airway with trickle of secretions or blood. In all the scenarios consent for emergency tracheostomy should al-

ways be obtained [7]. Various other options to access airway can be by placement of cricothyroid cannula with jet ventilation and nasal insufflation of oxygen with Total Intravenous Anaesthesia.

Progressive renal failure (in 75% of patients) can alter the metabolism and excretion. Drugs with active or toxic metabolites dependent on renal excretion including opioids like morphine, muscle relaxants like Vecuronium and Pancuronium and Sodium nitroprusside should be avoided [8]. Succinylcholine should be used sparingly in patients under treatment with cyclophosphamide because it inhibits pseudocholinesterase. NSAIDs should not be used. Atracurium and Cis-atracurium can be safely used for muscle relaxation. Among inhalational agents Desflurane or Isoflurane can be preferred.

In cases posted under regional anaesthesia, sensory neuropathy (10% of patients) if already present, should be documented preoperatively. Regional anaesthesia including peripheral nerve blocks can provide a suitable alternative in such patients for providing anaesthesia, immediate and post op pain relief wherever possible [9].

Prior to administering a regional technique, adequate coagulation status should always be determined [10].

Conclusion

Patient with Wegener's granulomatosis can always present with an undiagnosed difficult airway thus detailed airway examination and radiological assessment is a must. Multidisciplinary approach in the form of prior CO_2 ablation of subglottic stenosis, regional anaesthesia, Ultrasound-guided peripheral nerve blocks, TIVA can be used to avoid Cannot intubate situations.

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