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Case Report .

Wegener's Granulomatosis and Anaesthetic Implications: A Case Report

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ABSTRACT

Wegener's granulomatosis (WG), a rare immunopathologic disease, is a systemic vasculitis of small, medium and large arterial involvement. It presents a challenge to the anaesthesiologist due to multisystem involvement resulting in potential abnormalities of the airway, along with involvement of respiratory, circulatory, renal and central/peripheral nervous system. Additional morbidities arise in these patients secondary to treatment with corticosteroids and immunosuppressive drugs. A familiarity with proper approach to perioperative management is essential.

Keywords: Wegener's Granulomatosis, Vasculitis, Airway Lesion, Glomerulonephritis.

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INTRODUCTION

Wegener's granulomatosis (WG), is an uncommon immunopathological disease, characterised by a triad of necrotising granulomas in upper and lower respiratory tract, small and medium sized vessel vasculitis and glomerulonephritis. The vasculitis of veins, peripheral arteries, coronary arteries, granulomas and necrotizing changes are included in cardiovascular effects of WG.[1]

The first case was described by Heinz Klinger, a German medical student, in 1933. Three years later a German pathologist, Friedrich Wegener, described three additional cases and recognized the disorder as a distinct form of vasculitis in 1936. Other names occasionally used for Wegener's Granulomatosis are *Wegener's arteritis* or *Wegener's disease*. The estimated prevalence of 3 per 1,00,000 persons affected, makes this a very rare disease. Males and females are equally affected, whereas, it is more common in white population. The mean age of onset is approximately 40 years though it can be seen at any age.[2-4]

CASE REPORT

A 55 year old female, weighing 60 kg and 156 cm in height, a known case of WG since last 2 years, presented with complaints of pain in right side of mouth since 2-3 months. Pain was moderate in nature and relieved on taking medication. Patient had a history of dental extraction of upper right first molar, six months back. Post extraction she had complaints of nasal regurgitation of fluids that she was taking orally, for which she reported to dental OPD. On examination, a diagnosis of oro-antral fistula was made and surgical repair was done.

After that, patient was alright for a while but she started having pain on and off, in right side of her mouth about 2-3 months back for which she had been self-medicating herself until the pain became severe and she presented to the dental OPD again. She was advised surgical exploration in the right upper molar region for ulceration.

During preanaesthetic checkup, patient gave history of dyspnoea on routine household activities and occasional swelling of feet. There was no other significant past or present history. She underwent hysterectomy under subarachnoid block and cataract surgery for left eye under local anaesthesia three years back. Both of these surgeries were uneventful. On airway examination she had painful restriction of mouth opening to two fingers, due to previous surgery and ulceration. She had multiple missing teeth and neck movements were within normal range. Indirect laryngoscopy showed adequate chink of glottis.

On general physical examination, patient was moderately built, afebrile, heart rate-68/min; blood pressure-108/80 mm Hg and respiratory rate of 14 per min. The heart sounds were normal and chest was clear on bilateral auscultation. Her haematological profile showed haemoglobin-10.2 gm%, total leucocyte count-8500, platlets-2.4lac/cm^3, bleeding time-2'10", clotting time -4'40". Her metabolic panel revealed sodium of 138meq/L, potassium of 4.2meq/L, fasting blood sugar of 98mg/dL. Her blood urea was 22mg/dl and S. creatinine 0.7mg/dl.

Contrast enhanced computed tomography (CECT) of the chest revealed multiple tiny centrilobular nodules in both lung parenchyma predominantly in upper lobe. Linear fibrotic bands

were found in bilateral basal segments. She had t-wave inversion in leads v2-v6 on electrocardiogram. Echocardiography was done and patient had an ejection fraction of 30%, mild mitral regurgitation, with akinetic apex and decreased contractility at anterior septum and anterior wall at basal, mid and apical segment.

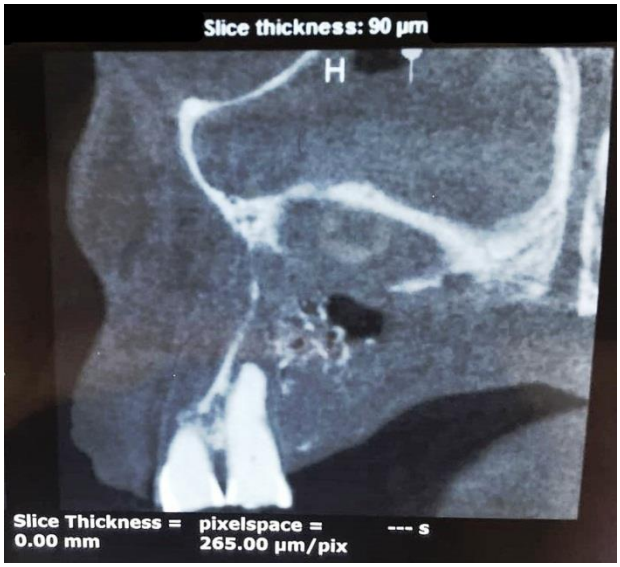

Fig 1: CT Maxilla

Patient was started on tab. Ecosprin 75 mg, tablet atorvastatin 10 mg, tablet ramipril once a day and tablet atenolol 50mg, twice a day. She was also taking tablet prednisone 10 mg since past 2 years since she was diagnosed with WG. Patient was posted in elective OT list for exploration and curettage under general anaesthesia.

Nil by mouth for 6 hours was ensured and written, informed high risk consent was obtained. On arrival in operation theatre all routine monitors were attached and baseline readings recorded. Peripheral intravenous (i.v.) access was secured with 18G cannula and ringer lactate started. Radial artery cannulation was performed next using ultrasound guidance, using out of plane approach and cannulation was achieved using Seldinger's technique. The arterial cannula was then used for invasive blood pressure monitoring in view of the compromised cardiac status. A difficult airway cart was kept ready for anticipated difficult airway in cases of Wegener's Granulomatosis. Injection glycopyrrolate 0.005 mg/kg i.v., Injection fentanyl 2µg/kg i.v. and injection hydrocortisone 100 mg i.v. were given. Patient was induced with injection propofol 1-2 mg/kg, using titrated boluses. Adequate bag mask ventilation was ensured and check laryngoscopy was done to assess the adequacy of airway and a Cormack- Lehane grading 2a was observed, as posterior portion of glottis was visible. Neuromuscular blockade was achieved with intravenous atracurium 0.5 mg/kg. Patient was ventilated for 3 min via facemask, using 2% sevoflurane and oxygen. A smaller size, cuffed endotracheal tube of 6.5mm internal diameter was used for endotracheal intubation, keeping in mind the probability of subglottic tracheal stenosis, under direct laryngoscopy and confirmed with chest auscultation and capnography. Patient was maintained with oxygen/nitrous oxide (50:50), sevoflurane and intermittent atracurium. Injection dexamethasone 8 mg i.v. was also given.

The surgery was uneventful. At the end of surgery all the inhalational agents were switched off and injection glycopyrrolate 0.01mg/kg and injection neostigmine 0.05mg/kg were given after confirming patient's spontaneous respiratory efforts. Endotracheal extubation was then successfully carried out once patient was awake and following commands.

DISCUSSION

Wegener's granulomatosis (WG) is a multisystem disorder of unknown aetiology. It is characterized by localized granulomatous inflammation of the upper and lower respiratory tract and systemic vasculitis involving small and medium-sized vessels associated with antineurotrophil cytoplasmic antibody (ANCA). The systemic vasculitic form of the disease can affect lungs and kidneys in the form of respiratory failure or necrotising glomerulonephritis.[3] A complete examination of both these systems including X-ray chest, CECT chest and renal function profile of the patient must be done in order to rule out their involvement.

Anaesthetic management in a case of WG depends upon the involvement of various organ systems as well as presence of difficult airway which may be due to involvement of upper respiratory tract. The mucosal lining of larynx may be replaced by granulation or fibrous tissue which may lead to narrowing of the lumen. This may be accompanied by destructive lesions of the epiglottis and narrowing of the subglottic space due to presence of proliferative lesions.[5] A preoperative indirect laryngoscopy must be done to rule out involvement of larynx and upper airway which could lead to narrowing of chink, which was normal in our patient.

A difficult airway must always be anticipated in this scenario and a difficult airway cart must be present in close vicinity in the operating room. It might include equipments such as different sizes of oral and nasopharyngeal airways, cuffed and uncuffed endotracheal tubes of smaller diameter, different sizes of both straight and curved laryngoscope blades, intubating laryngeal airway of variable sizes along with its endotracheal tubes, fiberoptic bronchoscope and finally all the instruments required for emergency cricothyroidotomy and tracheostomy.[6] Laryngoscopy and intubation must always be done gently and repeated attempts must be avoided as this can lead to bleeding from the granulomas or migration of the ulcerated tissue down into the trachea or larynx, causing airway compromise. In our patient we attempted a gentle check laryngoscopy to assess the airway as patient had painful restriction of mouth opening. Extra caution should be exercised in immediate postoperative period, after tracheal extubation, as these patients are more prone to developing airway oedema which may lead to airway obstruction. In our patient we administered injection hydrocortisone as these patients may have diminished stress response due to prolonged steroid intake as treatment for WG. Injection dexamethasone was also administered to decrease post-operative airway and oral oedema, as airway was a shared field in this case.

Anaesthetic drugs also need special attention in these patients as the drugs that require renal excretion may accumulate leading to adverse effects. The loading doses of inducing agents remain the same as they depend more on redistribution, rather than elimination. However, the maintenance doses of highly protein bound agents will be lowered by about 30-50 percent. Commonly used anaesthetic agents that depend predominantly on renal excretion are: vecuronium, pancuronium, atropine, glycopyrrolate

and neostigmine. Some drugs produce active or toxic metabolites that depend on renal excretion like midazolam, diazepam and morphine, hence should be used with caution.[7]

Cardiac involvement is seen in 6-25% of unselected patients and up to 44% of patients with severe renal involvement. Cardiac involvement was present in our case as evidenced by history of grade III dyspnoea and the echocardiography findings. A stable hemodynamic status needs to be maintained as variations in heart rate and blood pressure can lead to myocardial ischemia or infarction. Since the ejection fraction was severely reduced in our patient the doses of induction and maintenance agents were titrated in order to maintain a stable hemodynamic profile. Vasculitis of peripheral veins and arteries are included in cardiovascular effects of WG, hence number of arterial and venous punctures needs to be minimized, else patients may present with digital infarcts.[5] In our patient, due to the compromised cardiac status, invasive monitoring of blood pressure was warranted, so we performed ultrasonography guided radial artery cannulation, to confirm the patency of artery and to achieve cannulation in a single prick.

In 1966, Carrington and Liebow described 16 patients with pulmonary lesions identical to those of Wegener's granulomatosis with absence of or limited lesions elsewhere and, particularly, no focal glomerulitis.[8] Our patient also had no renal involvement and hence can be classified under limited form of Wegener's Granulomatosis. Lie JT proposed that the idea of 'limited form' of wegener's granulomatosis should probably be discarded as according to his observation , these patients may be in early or occult stage of the disease which may later and slowly present as full blown classical disease. So, ideally such patients should be advised and encouraged for regular follow up of their pulmonary and renal system.[9,10]

Even in cases where regional anaesthesia is an option, patients of WG may have preexisting neurological deficits, which need meticulous documentation. Another concern during regional anesthesia is increased risk of bleeding which may be due to the presence of circulating immune complexes leading to a low grade disseminated intravascular coagulation or as a result of complications from general vasculitis and granulomatous inflammation with cutaneous, meningeal, or spinal haemorrhages.[11]

In conclusion, Wegener's granulomatosis is a rare disease, hence not frequently encountered by the anaesthesiologists. It poses unique challenges demanding careful airway assessment and management. Proper planning and thorough preparation for both anticipated and unanticipated difficult airway, with proper documentation is the key to managing these cases.

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