Systemic amyloidosis: A challenge to the anaesthesiologists

Sir,

We present a case of a 66 year old male patient, a known case of systemic amyloidosis for tongue volume reduction surgery. Patient had a history of myocardial infarction, hypothyroidism, and hypertension which was controlled on medications. The recent myocardial ejection fraction was 45% with major adverse cardiac event (MACE) score of 1/6 with 6.6% calculated cardiac risk. Patient had history of obstructive sleep apnoea and the STOPBANG score was 7 out of 8. He underwent flexor retinaculum release surgery of left hand for carpal tunnel syndrome, 1 year back under supraclavicular block. The neck was woody in consistency in submandibular space. The inter-incisor distance was 3.5 cm, Mallampati grade IV [Figure 1], and restricted neck movements. Magnetic resonance imaging (MRI) face showed macroglossia with diffuse enlargement of tongue.

Antihypertensive and thyroid medications were continued till morning of surgery. Fasting protocol was followed. Standard American Society of Anesthesiologists (ASA) monitors were attached [Figure 2]. Radial artery cannulation was done before induction under local anaesthesia. Induction was done with etomidate (0.2–0.3 mg/kg) and muscle relaxation was facilitated with inj. succinylcholine 100 mg intravenous (IV), after pre-oxygenation for 3 min. In view of all signs of difficult intubation, we took the decision to use flexible fiberoptic-guided intubation. On first attempt endotracheal tube (ETT) 7.5 mm OD was threaded over fiberoptic bronchoscope (FOB) and with some difficulty we were able to negotiate the FOB but ETT was not negotiable over the FOB [Figure 3]. Subsequently, the FOB was removed and an ETT of 7.0 mm OD was threaded on it which was successfully placed in trachea. During intubation attempts, blood pressure dropped to 70/40 mmHg, which was unresponsive to mephentermine and fluid boluses, hence noradrenaline infusion was started. Anaesthesia was maintained as per standard technique. On completion of surgery, airway was secured with tracheostomy tube (7.0 mm ID Portex) in view of swelling of supraglottic structures in postoperative period. After return of consciousness and adequate spontaneous respiratory efforts, patient was shifted to HDU.

Amyloidosis is a rare disorder, characterised by extracellular deposition of abnormal protein fibrils in organs or tissues which progressively leads to organ dysfunction.[1] It may be inherited or acquired, localised or systemic, and life-threatening or an incidental finding. Amyloidosis disease is classified according to the type of amyloid protein as AL, AA, ATTR, Aβ2M, or Aβ.[2] AL type primary amyloidosis has clinical manifestations like cardiomyopathy, autonomic neuropathy, endocrine disorders, nephrotic syndrome, hepatosplenomegaly, and macroglossia.[3] Anaesthesia providers must be aware of

the multisystem involvement and their related anaesthetic implications. The anaesthetic concerns include airway difficulties, heart failure, arrhythmias, bleeding, and altered responses to pharmacological measures.

Amyloid deposition around head and neck with involvement in upper and lower respiratory tract may cause airway management difficulties. Macroglossia with poor mobility is a highly characteristic feature of AL amyloid, occurring in approximately 20%of the patients, and almost pathognomonic of systemic AL. Regional techniques should be considered over general anaesthesia wherever feasible, as regional blockade can obviate risks associated with airway management. Other advantages of regional anaesthesia include avoidance of polypharmacy, attenuation of the stress response, and superior analgesia.

The hypotension encountered on induction was resistant to conventionally used drugs. The haemodynamic instability could be due to an old myocardial infarction with septal wall dyskinesia and restrictive cardiomyopathy as well as autonomic dysfunction. Cardiac involvement represents the worst prognostic factor.[1] Heart failure is the most common terminal event for primary and hereditary amyloidosis. There are case reports mentioning cardiac adverse events like intraoperative death,[3] fatal perioperative myocardial infarction[4] and postoperative ventricular fibrillation.[5] Emergency drugs, defibrillators and pacers should be readily available due to high risk of cardiac arrhythmias and dysfunction. Isoflurane was used for maintenance because of its myocardial protective properties and lesser arrhythmogenicity.

Usually preoperative evaluation for causes of difficult airway beyond the vocal cords cannot be assessed. Such causes includes tracheal stenosis (polychondritis, sarcoidosis, papillomatosis, amyloidosis, or Wegener's granulomatosis, tracheobronchopathia osteochondroplastica[6], bacterial and fungal infections as well as tuberculosis), tracheal deviation, tracheal polyps, and tumours. Herein this case we encountered tracheal stenosis which necessitated us to intubate with smaller sized endotracheal tube.

Beside difficult intubation cart, preparedness with small size ETT is very important, because amyloid can get deposited below the vocal cords. The haemodynamic and cardiovascular instability necessitates meticulous care and preparedness with all resuscitation measures.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Figure 1: a : Photograph of patient showing thickening of periorbital, perioral and submental skin and subcutaneous tissue. Figure b is showing enlarged tongue with Mallampati grade IV

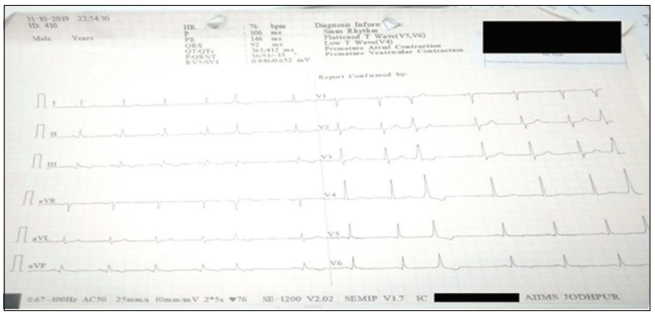

Figure 2: 12-lead electrocardiogram showing sinus rhythm with premature atrial contractions and T inversions in lead II, III, avf, V4, V5, V6

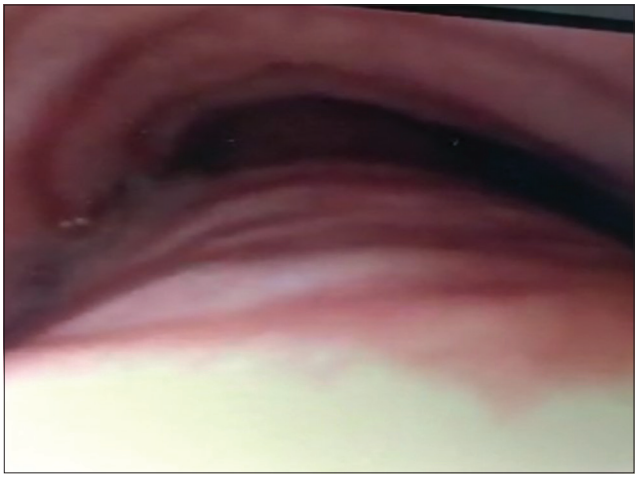

Figure 3: Bronchoscopic view of trachea showing amyloid invasion

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Conflicts of interest

There are no conflicts of interest.

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