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Case report

Anesthetic management of a case of retroperitoneal tumor with pemphigus vulgaris with multiple comorbid conditions

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Abstract We present a case of 50 year old female with a diagnosis of intraabdominal mass suffering from pemphigus vulgaris, myasthenia gravis, hypothyroidism, diabetes mellitus. Extra care to protect skin and mucus membranes from trauma during procedures is essential. Adrenocortical suppression following prolonged steroid therapy is another hazard to be tackled in the perioperative period.

1. Introduction

Autoimmune disease have important implications in anesthetic practice. Anesthesia for patients with multiple autoimmune diseases needs careful preoperative evaluation because of the diverse systemic effects of the disease. We present a case of 50 year old female with a diagnosis of intraabdominal mass suffering from pemphigus vulgaris, myasthenia gravis, hypothyroidism, diabetes mellitus. Extra care to protect skin and mucus membranes from trauma during procedures is essential. Adrenocortical suppression following prolonged steroid therapy is another hazard to be tackled in the perioperative period [1].

2. Case history

A 50 year old female presented with a history of drooping of both eyelids and diplopia 3 months back for which patient went to an ophthalmologist and was diagnosed as myasthenia gravis. Her serology at that time was positive for acetylcholine receptor antibodies. Following this patient developed multiple oral ulcers and raised red lesions followed by desquamation over chest, abdomen, back and extremities 2 months back and was diagnosed as pemphigus vulgaris. Patient was treated with pyridostigmine (30 mg thrice a day but discontinued for past 15 days without any clinical symptoms), immunosuppressants (azathioprine), steroids (prednisolone). Her other comorbidities included diabetes mellitus and hypothyroidism which were diagnosed a year back for which she was on oral hypoglycemic agents and thyroxine supplementation. Computerized axial tomography of the abdomen revealed a tumor of 5×4 cm in the subhepatic region encircling the superior mesenteric artery.

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At the time of preanesthetic evaluation the significant findings included the following: mouth opening of only 2–2.5 cm which was painful and Mallampatti grade III as tongue protrusion was difficult. Erosive oral mucositis grade 3 with lichenoid inflammation was bleeding to touch and the sputum was always stained with blood movements of cervical spine were normal. Skin lesions over extremities, chest and abdomen showed remission. Her current treatment included prednisolone 20 mg/day, OHA (Glimepride and Metformin), T. Eltroxin 50 mcg/day, T. Azathioprine 50 mg/day.

Routine investigations including hemogram, blood sugar, serum electrolytes, serum proteins and coagulation profile were within normal limits. Pulmonary function tests could not be adequately performed since the oral ulcers bled on touch.

With utmost care the cannulae were secured with 3M micropore since patient denied any fixation with sutures outside the operating room (OR). Patient was switched over to regular insulin along with 4 hourly blood sugar charting and dosage according to sliding scale. Insulin was given intravenously and subcutaneous insulin was avoided.

Patient was premedicated with intravenous sedative the night before and morning of surgery. Prokinetic agent (metoclopramide) and H_2 blocker (ranitidine) were given intravenously on the morning of surgery. No intramuscular premedications were given. Her routine dose of thyroxine 50mcg was given orally on the morning of surgery. Blood sugar, electrolytes and urine ketones were checked.

On arrival inside the OR the site of cannulation was checked for any signs of inflammation/eruptions. Noninvasive blood pressure (placed over cotton padding underneath), electrocardiogram and pulse oximetry (SpO2) were monitored. Preoperatively vital parameters were within normal limits. Anticipating a large surgical incision (and availability of lesion free lower back region) lumbar epidural catheter was placed. In anticipation of intraoperative bleeding (the tumor was vascular and encircling the superior mesenteric artery) the left radial artery, the basilic and the two peripheral veins were cannulated and secured with micropore. All these sites were followed till the end of case for any eruptions/rashes. Hydrocortisone 100 mg was given. Midazolam 2 mg and Fentanyl 100mcg were given for sedation and analgesia. Propofol 100 mg was given and Sevoflurane inhalation started. Ventilation was provided by applying minimal pressure on the face mask. Train of four (TOF) response was checked and found to be normal (TOF ratio > 0.9). Succinylcholine (SCh) was administered intravenously in a dose of 75 mg for neuromuscular blockade. Gentle airway instrumentation with lubricated laryngoscope was performed, glottis could be visualized, No. 7 Portex cuffed endotracheal tube inserted. Airway leak was obliterated by minimal inflation of the cuff. Throat pack soaked with saline adrenaline (1 in 200,000) was placed to control the bleeding and the tracheal tube was secured with a roller gauze bandage. Intraoperative bronchoscopy was done with utmost care to visualize the condition of tracheal mucosa. Tracheal mucosa was found to be inflamed and red down to the carina. We did not go past the carina to avoid any inadvertent injury (and its grave consequences) to the respiratory tree.

Anesthesia was maintained on Oxygen, Nitrous oxide, and Vecuronium (with neuromuscular monitoring). All vital parameters were well, maintained intraoperatively including the urine output. Steroid cover was given perioperatively. Surgery went on for 8hrs and blood loss was around 2.5L which was adequately replaced. Reversal was given (train of four response >0.9) but not extubated and shifted to ICU, sedated and electively ventilated overnight as the case finished by late evening.

Next morning extubation was planned. Patient was nebulized with levosalbutamol (0.63 mg) followed by adrenaline (1:10,000). Oral cavity was then packed with a fresh pack of adrenaline (1:2,00,000) and kept for 10 min. Repeat reversal with neostigmine 0.05 mg/kg and glycopyrrolate 0.01 mg/kg and patient was extubated after explanation of the complete procedure to the patient. Perioperative steroid was given as 100 mg/day for next 2 days and then tapered to routine oral dose. Blood sugar was maintained with gik infusion and sliding scale and thyroxine supplementation was continued in the postoperative period. All sites of cannulation were rechecked at regular intervals for any inflammation or bulla formation. Analgesia was maintained through iv and epidural routes.

3. Discussion

Presence of multiple autoimmune diseases complicates the anesthetic management as the considerations of each disease has to be taken into account. Primary considerations in this case are being the diseases itself and the various treatments received.

Pemphigus vulgaris is characterized by impaired cell adhesion within the epidermis leading to the formation of intra-epidermal blisters involving the skin and mucosa [2]. Nickolsky sign (lateral pressure causes separation of the epidermis) is a characteristic feature [3,4]. Prevention of friction and trauma is essential since either can lead to bullae formation. Use of adhesive tape is best avoided but due to lack of patient's consent and presence of healed skin surface single layer micropore was used for cannula fixation. Site of first iv cannulation was watched for inflammation/ bullous eruption. Lack of any eruptions in the first 24 h leads to fixation of other indwelling catheters (PICC, arterial line) with single layer micropore over healed skin surfaces. All these sites of fixation were carefully watched throughout the perioperative period till the time of discharge from hospital. Tracheal intubation is potentially dangerous in these patients in view of possible ulceration, bullae and edema formation around the glottic area and bleeding from preexisting lesions which can lead to life threatening airway obstruction [4]. Liberal lubrication of the laryngoscope, gentle laryngoscopy, use of a small tracheal tube (7 mm), adrenaline-soaked throat pack (which controlled bleeding) after intubation and before extubation, and avoidance of vigorous suctioning of oropharynx before extubation lead to an uneventful course. Neuraxial anesthesia and analgesia has been safely used for surgery in patients with pemphigus vulgaris [5,6]. We placed epidural catheter in lumbar region where the overlying skin was healthy and free of active/residual lesions. This site was also followed till epidural catheter was removed and thereafter till discharge from hospital. Management of these patients can be further complicated by drug therapy for pemphigus viz. preoperative steroid therapy produces hypothalamic pituitary- adrenocortical insufficiency. This requires perioperative steroid coverage to avoid Addisonian crisis. Other immunosuppressive drugs such as azathioprine and steroids predispose patient to infections,

bone marrow suppression and cardiorespiratory side effects [7,8]. This patient was given 100 mg/day of hydrocortisone for 3 days tapered over next 2 days to routine dose. Electrolyte imbalance, dehydration and hypoalbuminemia are present in 5–6% of cases of pemphigus [9]. Adequate fluid and electrolyte replacement was maintained perioperatively.

Myasthenia gravis (MG) is the prototype of antibody mediated autoimmune disease. Myasthenia gravis results from the production of autoantibodies against the acetylcholine receptor (AChR) of the neuromuscular synapse. Repetitive nerve stimulation results in a decremental response. The disease is frequently associated with morphological abnormalities of the thymus. Medical treatment consists of anticholinesterases, corticosteroids and immunosuppressants [10]. Abnormalities of thymus were ruled out in the CT scan findings of this patient and her pyridostigmine was stopped 15 days back and she was clinically asymptomatic for MG. The most sensitive diagnostic criterion of myasthenia gravis is the decrement in the muscular response to repetitive supramaximal stimulation of a peripheral nerve. Mann et al. concluded that myasthenic patients with a preanesthetic fading after train-of-four (TOF) stimulation have a significantly decreased ED95 of atracurium that is consistent with an increased sensitivity to nondepolarizing neuromuscular blocking agents in myasthenia gravis patients whereas myasthenic patients without fading had a mean ED95 value of 0.24 mg/kg atracurium, which is similar to the ED95 of nonmyasthenic patients [11]. We measured the preoperative TOF response of this patient under inhalational anesthesia and adequate analgesia (TOF ratio > 0.9) before giving SCh. Further doses of NMBA were guided by TOF monitoring.

Patient received her routine dose of thyroxine supplementation on the morning of surgery and a careful watch for arrhythmias was maintained. Normothermia was maintained and nasogastric tube was placed for gastric emptying. Blood sugar was controlled perioperatively with GIK (Glucose, insulin and potassium) infusion and frequent blood sugar monitoring. This case report reaffirms the notion that anesthesia is an anticipatory science and with increased vigilance and careful planning, patients with multiple autoimmune diseases may have an uneventful perioperative course.

Conflict of interest

There is no conflict of interest.

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