Perioperative anesthetic challenges in Alkaptonuria patient with comorbid conditions

Dear Editor,

Alkaptonuria (AKU) is a rare autosomal-recessive inborn error of tyrosine metabolism which is a progressive debilitating disease and symptomatic around 30 years of age. In AKU, patient lacks enzyme required to breakdown homogentisic acid and on exposure to air complains of "black nappies." Deposition of the onchronosis pigment in the large joints and spine causes joint destruction. In addition, there is involvement of airway, respiratory, renal, cardiac and coagulation system.[1]

A 56-year-old man with AKU, G6PD deficiency, hypothyroidism, and hypertension, was scheduled for unilateral total knee arthroplasty. He was receiving levo-thyroxine 50 μg and amlodipine 5 mg once a day (OD). Over a span of 13 years the patient underwent multiple surgeries including lumbar thoracic, cervical fixation and, open cholecystectomy under general anesthesia. Neurological examination revealed hyperreflexia and spasticity in the lower limbs with decreased sensation over S1 dermatome. Airway examination revealed mouth opening of 5 cm, thyromental distance of 5.5 cm, limited neck extension (<10°), and a Mallampati class IV. Rest all investigations were normal. Patient was kept nil per oral prior to surgery and amlodipine and levo-thyroxine on the day of surgery.

In the operating theatre, an 18 G intravenous line was secured, mandatory monitoring was attached and a difficult airway cart was kept ready. Patient was asked to self-position on the operation table. Following pre-oxygenation, IV fentanyl 2 μg/kg and propofol 2 mg/kg were administered. After loss of eyelash reflex, a check laryngoscopy was done with video-laryngoscope C-MAC® (Karl Storz, Germany) using a D-Blade which revealed a POGO of 75% with a granular deposit over the left posterior arytenoid, which adequate glottic chink. Vecuronium bromide 0.1 mg/kg was administered and trachea was intubated with 8.5 mm ID. Anesthesia was maintained with nitrous in oxygen, propofol infusion at 50-100 μg/kg/min, and intermittent boluses of vecuronium. At the end of surgery, an ultrasound-guided continuous femoral nerve block with 0.2% ropivacaine 15 ml bolus was performed followed by infusion of ropivacaine 0.2% with fentanyl 2 μg/ml. The surgery lasted for 2 h and vitals were stable during the entire period. 1.5 L of crystalloids were infused. Muscle paralysis was reversed. Black discolouration of heat and moisture exchanger as seen in this patient [Figure 1]. Trachea was extubated and the patient had an uneventful postoperative period.

Caring for a patient with AKU ochronosis with multiple comorbidities is challenging. Deposition of HGA in the cartilage of the airway and respiratory system can result in glottic stenosis leading to difficult airway management.[2] HGA deposits also make the dura and the arachnoid membrane vulnerable, therefore central neuraxial techniques were not preferred in the present case.[3] Drug-related precautions were observed for G6PD deficiency.[4]

Figure 1: Black discolouration of heat and moisture exchanger in this patient

The learning points in this case is the preparedness for anticipated difficult airway, multimodal analgesia and good coordination with orthopedic team for perioperative management. Use of VL with shorter learning curve has changed the management in patients with difficult airway.[5,6] Since there is no approved treatment for AKU, with every repeated surgery these patients have progressively increasing anaesthetic challenges necessitating meticulous anesthetic management.

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There are no conflicts of interest.

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