Anesthetic management in a patient with Kindler’s syndrome

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ABSTRACT
A 35-year-old male with pan-anterior urethral stricture was scheduled to undergo perineal urethrostomy. He was a known case of Kindler’s syndrome since infancy. He was having a history of blister formation, extensive poikiloderma and progressive cutaneous atrophy since childhood. He had a tendency of trauma-induced blisters with clear or hemorrhagic contents that healed with scarring. The fingers were sclerodermiform with dystrophic nails and inability to completely clench the fist. Airway examination revealed thyromental distance of 7 cm with limited neck extension, limited mouth opening and mallampatti class III with a fixed large tongue. He was reported as grade IV Cormack and Lehane laryngoscopic on previous anesthesia exposure. We described the anesthetic management of such case on guidelines for epidermolysis bullosa. In the operating room, an 18-G cannula was secured in the right upper limb using Coban™ Wrap. The T-piece of the cannula was than inserted into the slit and the tape was wrapped around the extremity. The ECG electrodes were placed on the limbs and fixed with Coban™. Noninvasive blood pressure cuff was applied over the wrap after wrapping the arm with Webril® cotton. Oral fiberoptic tracheal intubation was done after lubricating the laryngoscope generously with a water-based lubricant with 7-mm endotracheal tube. Surgery proceeded without any complication. After reversing the residual neuromuscular block, trachea was extubated once the patient became awake. He was kept in the postanesthesia care unit for 2 hours and then shifted to urology ward.

Key words: Difficult airway, epidermolysis bullosa, Kindler’s syndrome

INTRODUCTION
Kindler’s syndrome (KS) is a rare autosomal recessive genodermatosis, which presents itself in infancy.[1] The syndrome is a combination of features of inherited blistering skin disorders [e.g., dystrophic epidermolysis bullosa (EB)] and congenital poikilodermas (e.g., Rothmund-Thompson syndrome).[2] Unlike EB, the literature on the anesthetic and airway management of patients with KS is limited. We described the anesthetic management of a rare case of KS scheduled to undergo perineal urethrostomy that was managed on the guidelines for EB.

CASE REPORT
A 35-year-old male with pan-anterior urethral stricture was scheduled to undergo perineal urethrostomy. He was referred to the anesthesia department for preanesthetic evaluation. He was a diagnosed case of Kindler’s syndrome with a history of blister formation, extensive poikiloderma and progressive cutaneous atrophy since childhood. Although, one sibling and both the parents were normal, one of the siblings has the same skin disorder. Blistering tendency was noted immediately after delivery. He had a tendency of trauma-induced blisters with clear or hemorrhagic contents that healed with scarring. The blistering tendency gradually subsided by the age of 19 years. Reticular erythema was noticed first on the face, affecting the forehead and cheeks in infancy; spreading progressively to the neck and the chest, and later to all over the body. Increased photosensitivity with sunburn after minimal sun exposure was present from early infancy.

His only anesthetic exposure was for phimosis at 16 years of age. For the planned procedure, the anesthetic management was on the guidelines for epidermolysis bullosa. In the operating room, an 18-G cannula was secured in the right upper limb using Coban™ Wrap. The T-piece of the cannula was then inserted into the slit and the tape was wrapped around the extremity. The ECG electrodes were placed on the limbs and fixed with Coban™. Noninvasive blood pressure cuff was applied over the wrap after wrapping the arm with Webril® cotton. Oral fiberoptic tracheal intubation was done after lubricating the laryngoscope generously with a water-based lubricant with 7-mm endotracheal tube. Surgery proceeded without any complication. After reversing the residual neuromuscular block, trachea was extubated once the patient became awake. He was kept in the postanesthesia care unit for 2 hours and then shifted to urology ward.

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years of age. He was reported to be a difficult airway with grade IV Cormack and Lehane laryngoscopic view on direct laryngoscopy and the airway has been secured using a fiberoptic bronchoscope after single failed attempt with Macintosh laryngoscope. The physical examination revealed reticular skin patches, erythema and telangiectasiae all over the face, neck and the upper parts of the chest. Cigarette-paper like wrinkling of the skin on the abdomen, hands and feet was present [Figure 1a]. The fingers were sclerodermiform with dystrophic nails and inability to completely clench the fist [Figure 1b]. Cardiovascular and respiratory systems were normal. Airway examination revealed a thyromental distance of 7 cm with limited neck extension. His mouth opening was 2.5 cm, [Figure 1c]. He was mallampatti class III with a fixed large tongue. His lower incisors were missing and upper three teeth were artificial and fixed [Figure 1c]. Laboratory investigations revealed normal hemogram, coagulogram, serum electrolytes. Renal and liver function test were within normal limits. In view of the predicted difficult airway, awake fiberoptic intubation was planned.

The patient was premedicated with oral ranitidine 150 mg and oral alprazolam 0.5 mg a night before and on the morning of surgery. An 18-G venous cannula was secured in the right upper limb using 3M™ Coban™ Self-Adherent Wrap. A small slit was made in a 6-inch piece of Coban™, 1 inch from the end of the tape. The T-piece of the cannula was than inserted into the slit and the tape was wrapped around the extremity. The tubing of the intravenous drip-set was further included in the Coban™ wrap. Intravenous glycopyrrolate 0.2 mg was administered as antisialogouge and patient was nebulized with 4 ml of 4% lignocaine.

In the operating room, standard 5-lead electrocardiogram (ECG) was applied after trimming the adhesive from the adult ECG electrodes. The electrodes were then placed on the limbs and fixed to the skin by wrapping the limbs with Coban™. After wrapping the left arm with Webril® cotton, noninvasive blood pressure (NIBP) cuff was applied over the wrap. Adult clip-on pulse oximetry probe was applied over the right index figure. Fiberoptic laryngoscope was lubricated generously with a water-based lubricant (KY® jelly) and loaded with 7-mm endotracheal tube (ETT). Oral fiberoptic intubation was performed using spray-as-you-go technique. The ETT was secured without tape by snugly tying a surgical mask around the back of the head. Anesthesia was then induced with fentanyl 100 mcg, propofol 140 mg and 5 mg vecuronium was given. The patient was maintained on propofol infusion and oxygen and nitrous oxide. One milligram vecuronium was repeated every 20-25 minutes. Surgery proceeded eventufully and lasted for 190 minutes; on completion of surgery paracetamol infusion was started and residual neuromuscular block was reversed with neostigmine 3.5 mg and glycopyrrolate 0.7 mg. Trachea was extubated once the patient became awake. He was kept in the post anesthesia care unit for 2 hours and then shifted to urology ward.

DISCUSSION

KS, first described in 1954 by Theresa Kindler is a very rare (1 in 100000) hereditary disorder characterized by acral blister formation in infancy and childhood, progressive poikiloderma, cutaneous atrophy and increased photosensitivity.[1]

The patients with KS may present for plastic procedures to correct pseudosyndactyly of the hands or feet and to increase oral opening, esophageal dilatation and gastrostomy, dental restoration, procedures for urethral or anal stenosis. Several precautions and attention should be paid to anesthetize such a case. Premedication with sedative drugs should be given in younger patients to avoid restlessness and trauma, particularly when inhalational induction is planned. Larger doses are usually required. Adult patient should be brought in fully awake condition so as to assist in proper positioning.[8]

In the intraoperative period, routine anesthetic monitoring includes NIBP, ECG, pulse oximetry and core temperature. Further monitoring should be limited to the minimum, according to the surgical procedure that is going to be carried out and the risk associated to the patient. Electrocardiography electrodes can cause trauma during application and removal. Various methods of ECG electrodes placement have been described. Electrodes can be freed from adhesive part and electrodes can be directly laid beneath the patient[6] or wrapped in mummified thoracic dressing for good contact.[9] Adhesive ECG electrodes can also be placed on undersurface of a chest radiograph over

Figure 1: The patient of Kindler’s syndrome (a) cigarette-paper-like wrinkling of the skin on the abdomen, (b) finger deformity with dystrophic, (c) limited mouth opening on airway examination
suitably placed holes; the patient is then placed over the film. Needle ECG electrodes can also be used. Digital pulse oximetry probe is mostly difficult in these patients due to joint deformity and associated syndactyly, ear probe may be more useful. For monitoring of the NIBP it is necessary to place several soft nonadherent gauzes under the cuff. Invasive blood pressure monitoring is recommended for lengthy procedures or wherever indicated but scarring over wrist may make radial artery cannulation difficult. Lubricated axillary probe should be used for temperature monitoring.

The protection of all pressure points in the skin is essential. Careful lubrication and padding of contact surface to reduce shearing force and epithelial damage should be considered. Eyes should be protected with care. Simple eye ointments should be used abundantly after anesthetizing the patient. Special attention to eyes should be paid if the surgery should be done in prone position.

Adhesive tape used to secure the intravenous catheters, ETTs and other monitoring devices may cause skin damage. Sutures and roll of the gauzes should be used to secure devices. The intravenous catheter should be fixed with nastro, vaselinated gauze or line of suture.

General anesthesia is the technique of choice with a low incidence of airway blisters formation. Intravenous induction is generally less traumatic than mask induction and propofol is a better choice over thiopental as it provides smoother induction. However, intravenous assess may be difficult due to atrophied and fragile skin. Inhalational induction may then become necessary.

Airway issue is a main concern in these patients. Repeated scarring from healed bulla on skin and mucosal surface can result in reduce mobility of joints and narrowed mouth opening. Individual affected with this condition have limited mouth opening, ankyloglossia, dental overbite and atrophy of buccal mucosa. Difficult airway management is expected in such a case. Bullae in the oropharynx and hypopharynx may rupture with spillage of serosanguinous fluid into the airway. Contracture of other joints makes oropharynx and hypopharynx cause difficult airway. Application of facemask may be difficult and may cause trauma and blisters if tightly applied. Facemask can be applied with several layers of vaseline gauze beneath. Fiberoptic intubation causes lesser trauma to the mucosal epithelium than insertion of a laryngoscope, and should often be the first choice. Oral intubation should be preferred over the nasal intubation as bullae may be induced at the opening of the nares by a tight ETT. Suctioning of oropharynx should be avoided to reduce the risk of big and hemorrhagic blisters formation.

The regional anesthesia is an alternative to the general anesthesia because there is no need of airway manipulation but there is controversies regarding use of regional anesthesia with predicted difficult airway, “New Thoughts and Concepts” published by Benumof in ASA refresher course book stated that use of regional anesthesia in a patient with known difficult airway require high degree of judgment and concludes that it is unacceptable to do regional anesthesia with a known difficult airway when surgery cannot be terminated early or there is poor access to patients head. Beneditto stated that an anticipated difficult airway is not a contraindication to the use of regional anesthesia techniques. Optimum anesthesia management mandates individualized assessment of the risk/benefit balance. Also blisters, trauma and infection at sites of puncture contraindicates regional anesthesia. The subcutaneous infiltration with a local anesthetic should be avoided due to the possibility of epidermis dissection and formation of big blisters. After puncture, the area should be covered with lubricated gauze.

In postanesthesia care units the utilization of oxygen rich atmosphere should be preferred instead of facemask and nasal probe. There is a risk of airway obstruction due to big blister or hematoma formation in oropharynx or hypopharynx. Careful monitoring should be paid in postoperative period for early detection of airway obstruction. Proper analgesia should be given in postoperative period to avoid agitation and iatrogenic trauma.

In conclusion, we described the safe conduct of anesthesia in an adult patient of KS with airway difficulty. We recommend that these patients should be managed on the guidelines for the anesthetic management of EB; a closely related but more common disorder than KS.

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