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General anesthesia in a patient with dystrophic epidermolysis bullosa

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Abstract
A 13-year-old boy with epidermolysis bullosa underwent a repair of pseudosyndactyly. He had a long history of bullae formation in the oral cavity and on the pharynx and body surface, and some were active at the time of surgery. We chose inhalational general anesthesia with tracheal intubation using sevoflurane and nitrous oxide. The trachea was successfully extubated after the surgery, and no major bulla formation was observed. General anesthesia with tracheal intubation may be chosen as anesthesia for patients with epidermolysis bullosa.

Key words Epidermolysis bullosa · Anesthesia · Children · Intubation

Introduction

Distrophic epidermolysis bullosa (DEB) is an autosomal recessive disorder, which is characterized by cutaneous blistering and subsequent scarring caused by relatively minor stimuli or friction. Great care should be taken with the anesthetic management of patients with this rare disease. Avoiding the formation of new bullae during perioperative management is always a challenge for anesthesiologists. In particular, bulla fomation in the airway must be avoided. We report a patient with severe DEB who underwent a repair of pseudosyndactyly under general anesthesia.

Case report

A 13-year-old boy (125 cm, 20 kg) presented for elective surgical correction of pseudosyndactyly on both hands. His medical record revealed that he had been diagnosed as having DEB soon after his birth. At the age of a few months, he had surgery for inguinal hernia under inhalational general anesthesia via face mask without any major problem perioperatively. He had a history of oral, pharyngeal, and esophageal blistering, occasional ulcer formation, and poor dentition; consequently, his mandibular movement was limited, and he had eating difficulties. He had experienced repeated blistering all over his body with rupturing and scarring. He had active blisters on his back, chest, and shoulder at the time of admission to our hospital. All his fingers and toes were fused into lumps because of repeated rupturing of blisters and scarring. Routine preoperative laboratory testing revealed that hemoglobin was 5.5 $g \cdot dl^{-1}$ and albumin was 1.93 $g \cdot dl^{-1}$. An electrocardiogram (ECG) showed tachycardia and left ventricular hypertrophy, and echocardiography showed mild enlargement of the ventricle cavity with normal wall movement of both ventricles.

Eight milligrams of oral midazolam was given 30 min prior to anesthesia because he was anxious. A fiberoptic bronchoscope and laryngeal mask airway were prepared for a possibly difficult intubation. We monitored his ECG, pulseoxymetry (Sp_{O_2}), and capnograph during the procedure. ECG was monitored with the use of the usual adhesive dots, but Vaseline was placed between his body surface and the dots. A nonadhesive Sp_{O_2} probe was attached to his right ear. Display of the monitoring data of both ECG and Sp_{O_2} were excellent during the operative period. Neither noninvasive blood pressure measurement with cuff nor intraarterial blood pressure monitoring was carried out because they could cause new bulla formation and because only a minor procedure was scheduled. General anesthesia was induced with 50% nitrous oxide and 5% sevoflurane with oxygen via a soft-type face mask, which was well lubricated with Vaseline. An intravenous access was placed in the right cubital fossa and fixed by suture. Forty micrograms of fentanyl and 2.5 mg of vecuronium were ad-

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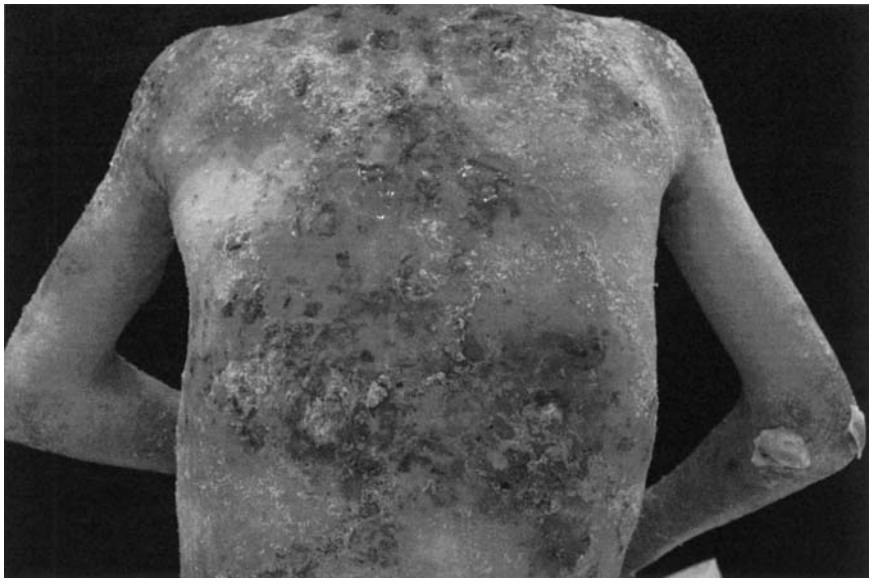

Fig. 1. Preoperative appearance of bullae and blistering on the back of a patient with epidermolysis bullosa

ministered after control ventilation via a face mask was established. Although his mouth opening was limited, the vocal cords were visualized with a laryngoscope well lubricated with Vaseline. The trachea was intubated with a 5.5-mm uncuffed endotracheal tube, which was also lubricated with Vaseline. Although there was minor leakage around the tracheal tube, it was acceptable for positive pressure ventilation. The tube was fixed by a wide string tied around his neck and also lubricated with Vaseline. Surgery involved the release of syndactyly of both hands and a skin graft from his thigh. This procedure was scheduled to last a maxmum of 2 h, but it took 5 h. Red blood cells were given during surgery because of anemia, although the total blood loss was only 20 g. His heart rate did not change throughout the procedure. The trachea was extubated after leakage around the endotracheal tube was confirmed, but there was no airway problem. New minor bulla formation was seen around his mouth, which was likely due to face-mask manipulation during the induction of anesthesia, but no new bullae were observed at ECG dot sites or the S_{PO_2} site. The patient was observed overnight in the intensive care unit (ICU), and he was discharged from the ICU the next day with no major problems.

Discussion

No complications occurred during or after general anesthesia with tracheal intubation in a patient with DEB. The choice of anesthetic methods was the main issue. First, regional anesthesia could have been chosen to manage this case. Diwan et al. [1] reported that they successfully managed a 4-year-old patient with a continuous axillary block. However, they also gave small amounts of intravenous ketamine, inhalational nitrous oxide, and 0.5% halothane via head box. Kelly and colleagues [2] also reported successful brachial plexus anesthesia in patients with DEB. Second, total intravenous anesthesia with spontaneous breathing preserved was another possible choice. Griffin and Mayou [3] reported that only 7% of 469 surgical patients with DEB received intravenous ketamine anesthesia and 3% received propofol. However, they did not describe the anesthetic methods in detail. Lin and colleagues [4] reported the use of ketamine in 11 out of 129 episodes anesthesia and propofol in 6 episodes. One of the 11 patients who received ketamine anesthesia required an oral airway.

We chose general anesthesia for our patient for several reasons. First, this operation was scheduled to last 2 to 3 h, and it involved both hands and a skin graft. It would have been necessary to administer a reasonable amount of local anesthetics to complete this surgery if the brachial block technique had been chosen for both hands. Moreover, our patient was a 13-year-old boy who was very anxious at the time of the preoperative visit. Second, although intravenous anesthesia may reduce new bulla formation in the airway, there is a risk of airway compromise. Furthermore, if an airway compromise should occur it would create an urgent situation in a patient in whom airway insertion or intubation might be difficult because of the limitation of his jaw movement.

In two reviews [3,5], the rate of new bulla formation associated with intubation was reported to be very rare, and bulla formation occurred only after dental or esophageal procedures [5], mainly in association with

difficult intubation [3]. Moreover, because the trachea is composed of epithelial column cells, whereas the pharynx and larynx are constructed from squamous cells, if the laryngoscope and tracheal tube are well lubricated before use, tracheal intubation and tracheostomy should be a relatively safe procedures [6]. A laryngeal mask airway is another technique to be considered. However, its use in patients with DEB has not been widely described in the literature. Ames et al. [7] reported it to be safe in patients with epidermolysis bullosa, although in one patient a single new lingual bulla formed. The present patient had a history of multiple episodes of oral bulla formation with rupturing and scarring; thus, we considered tracheal intubation a safer method of airway management than the use of a laryngeal mask airway.

In conclusion, we successfully managed the anesthesia in a patient with a severe case of DEB. General anesthesia with tracheal intubation is an appropriate choice of anesthesia in a patient with DEB.

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