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Case Report Open Access

Anesthetic Considerations in Pediatric Patients with Epidermolysis Bullosa: A Case Report

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Abstract

Epidermolysis bullosa (EB), a rare genetic disorder with abnormal fragility of skin and mucous membranes, is characterized by blister formation, either spontaneously or following a minor friction. In addition to the skin, it often involves other epithelial-lined organs requiring profound considerations in the anesthetic management related to some comorbidities. Monitoring, difficult airway, positioning, nutritional deficiencies and poor immunity can be challenging for the anesthesiologist in perioperative management. The authers described an experience of a 3-year-old male child with a dystrophic type of EB since birth who underwent hernioplasty. Perioperative careful management and minimal mandatory monitoring might be needed to minimize mechanical injury to the skin and mucous membranes.

Keywords: Epidermolysis bullosa; Pediatrics; General anesthesia

Introduction

Epidermolysis bullosa (EB) is an uncommon hereditary skin disorder characterized by abnormal fragility of the skin and mucosal surface. Vesiculobullous lesions may appear following a trauma, exposure to heat or even for no obvious reason. The pathophysiology of EB depends on the contributing defect in the epithelial or subepithelial connective tissue by application of friction or shearing forces, with varying degrees of severity from blister formation to life-threatening medical conditions [1,2].

EB has more than 20 different described subtypes that may have either localized or generalized dermatological manifestations [3]. Although more than 20 clinical subtypes have been described, usually 3 major categories are classified as EB simplex, junctional EB, and dystrophic EB according to the involvement of dermal layer in blister formation. EB simplex is the most common type that has intraepidermal blistering and minimal mucosal involvement. Junctional EB, the least common form, is diagnosed when blistering or separation occurs between the epidermis and dermis. Dystrophic EB, on the other hand, involves the level of the dermis [4].

In addition to considerations associated with the skin and mucous membrane susceptibility, anesthetic management of EB should focus on the airway involvement of disease. Most EB involves oral and pharyngeal mucous membranes. Moreover, some types of EB additionally involve the tracheal epithelium. Safe anesthetic management requires careful anesthetic planning and preparation for airway management, as well as clinical monitoring and minimal touch.

There were few cases reported on general anesthesia with anesthetic management of patient with EB. Thus, we report presented the successful anesthetic management of a pediatric patient with EB undergoing inguinal hernioplasty.

Case Report

A 40-month-old male child with known dystrophic EB since birth was admitted for sustained projectile vomiting and abdominal distension. The diagnosis was incarcerated inguinal hernia and he had to undergo hernioplasty because manual reduction was failed.

The patient was born with normal delivery at 38 weeks, weighing 2.58 kg. Since birth, he had presented multiple eruptions and blisters on the entire body with no other perinatal problems. He therefore required frequent hospitalization for several episodes of infection, sepsis, anemia and incarcerated hernia.

On physical examination, the patient weighed 8.1 kg with a height of 74.3 cm, presenting growth retardation due to poor oral intake from oral ulceration. He had numerous blisters and generalized multiple scars, eruption and bullae on the body. Moreover, the joint of extremities were contractured, the skin over the hand and feet were highly atrophic, and the fingernails and the toenails were absent (Figure 1). He had few teeth with caries and dysplasia. Preoperatively, the patient was performed airway examination including the amount of mouth opening and tongue attachment to the mouth floor. Limitation of neck extension was not found. However, the involvement of pharynx or trachea could not be visually confirmed. The patient's cheek and perioral area that were involved and injured were covered with atraumatic foam dressing pads for skin protection.

In the operating room, standard monitoring using pulse oximetry, electrocardiogram, and noninvasive blood pressure were measured during the operation. To prevent the injury of skin, the sensor of pulse oximetry was applied using a minimal adhesive portion, and a cotton wrap was laid under the blood pressure measuring cuff. On electrocardiogram monitoring, the adhesive part was cut off leaving only the gel portion of the pad. The atraumatic foam dressing pads were also helpful to lubricate and stabilize any other anesthetic monitoring devices (Figure 2).

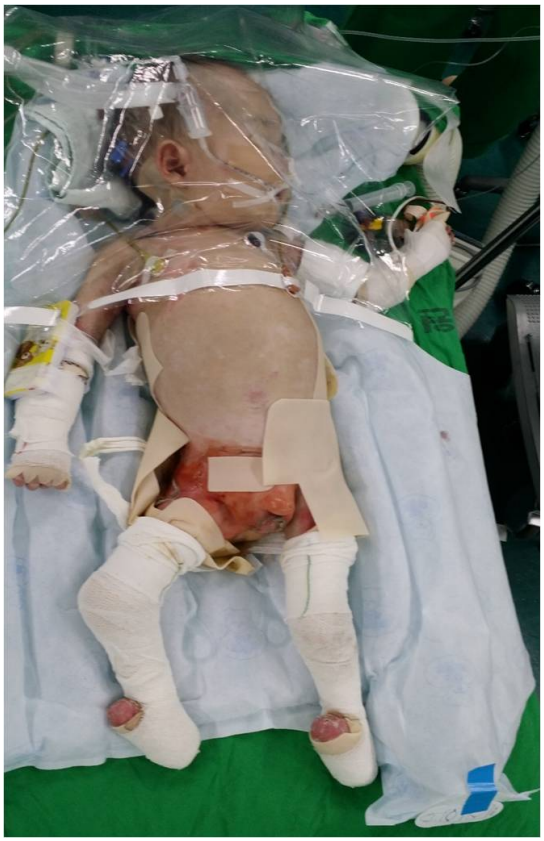

Figure 1: Overall view of patient's skin manifestations and protective application of anesthetic monitoring.

In preparation for the risk of difficult airway and traumatic intubation, C-MAC® video laryngoscope (Karl Storz Endoscopy, Tuttlingen, Germany) was prepared in addition to conventional equipment such as masks, endotracheal tubes and stylets. Furthermore, readiness for tracheostomy was maintained for an emergency airway situation.

After preoxygenation, anesthesia was induced with intravenous ketamine 15 mg and glycopyrrolate 0.1 mg along with oxygen and sevoflurane under gentle mask holding. Rocuronium 10 mg was administered after confirming ventilation. Minimal pressure for chin lift and head tilt was used for manual ventilation. Endotracheal intubation was successfully achieved using C-MAC® video laryngoscope with a 3.5 cuffed tube, and the endotracheal tube and blade were also lubricated. Fortunately, the oropharyngeal airway was intact permitting successful endotracheal intubation. For minimizing trial of intubation, we used a cuffed endotracheal tube. The cuff on the endotracheal tube was not inflated, and the tube was fixed with adhesive bandage over the protecting pad on the skin. Since the patient had very poor peripheral intravenous access and needed postoperative TPN nutrition, a central catheterization was performed on the right internal jugular vein under ultrasound guidance. The catheter was secured with tagging-suture instead of protecting bandage. In addition, ophthalmic ointment was applied on the eyes. During the entire process, the patient was manipulated with sterile gloves.

Anesthesia was maintained with 2.0-3.0 vol % sevoflurane and 50% oxygen and medical air. Large amounts of fluids including crystalloid and colloids were needed to maintain adequate perfusion during the operation. But no transfusion was needed. At the end of the operation, neuromuscular blockade was antagonized, and tracheal extubation was done without problems including airway obstruction, spasm or excessive secretion.

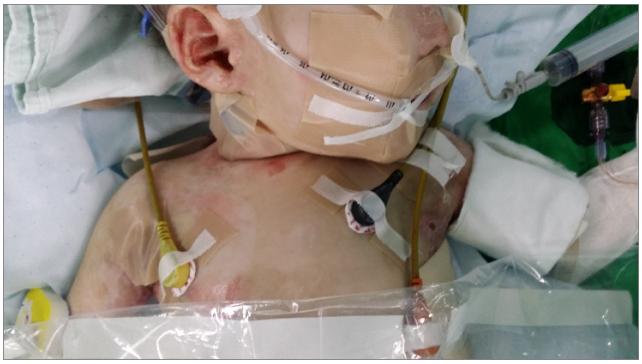

Figure 2: Securing endotracheal tube, central catheter and other anesthetic monitoring.

In the post anesthetic care unit, the patient showed stable vital signs. The pulse oximetry was monitored continuously and SpO_2 was maintained above 98% without oxygen supply. Postoperative analgesia with fentanyl 1.25 mcg/kg/hr intravenously provided adequate pain relief. The patient was sent to general ward and discharged without other complications 12 days after the operation.

Discussion

In this report, the patient was dystrophic EB. This type, with a prevalence of about 2 in 100,000 [5], is characterized by the presence of extremely fragile skin and painful blister formations in the skin owing to the loss or absence of normal intracellular bridges in response to minor trauma, friction, or pressure. This results in scar formation and repetition of injury and healing. Although some are confined to the skin, more severe forms also involve other epithelial-lined organs, including the oral cavity, external eyes, and gastrointestinal and genitourinary tracts, as well as the bone marrow and musculoskeletal system [6]. Therefore, anesthetic management according to the comorbidity is important and challenging to the anesthesiologists.

Anesthesia in patients with EB presents considerable problems. Their energy requirements are high due to injury and healing of skin, while oral intakes are decreased as a result of oropharyngeal and esophageal lesion. Moreover, absorption is decreased due to gastrointestinal lesions. For these reasons, they often suffer from growth retardation. Oral scarring with limited mouth opening, esophageal stricture, anemia, and infection are also common. Nutritional deficiency leads to hypoproteinemia, anemia and electrolyte imbalance that may affect the pharmacological effects of anesthetic agents [7]. Acquired syndactyly in hands and feet following skin contracture may lead to loss of fingers and toenails [8]. Infection is also common because of decreased immunity. Dental problems are frequently associated including enamel dysplasia, defective crowns, severe caries, loose teeth and cleaning difficulty [9,10].

Fortunately, most EB including simplex and dystrophic types do not have tracheal involvement. However, the junctional type of EB often has laryngeal and tracheal lesions because the trachea has pseudostratified, columnar, ciliated epithelium, whereas the oral, pharyngeal and esophageal mucosa is lined with stratified squamous epithelium [11]. Airway complications such as edema and obstruction due to blister and scars can occur after extubation and emergent tracheostomy should be kept in readiness. Otherwise, mask ventilation without tracheal intubation should be considered. Since our patient had dystrophic type of EB we did not expect tracheal and laryngeal involvement. Several authors reported successful tracheal intubation without any other specific event or complication in patients with dystrophic type of EB [12,13]. Therefore, we conducted general anesthesia with tracheal intubation.

Although laryngeal and tracheal involvement of EB is rare [12], the involvement of the skin and mucous membranes of the oral cavity and oropharynx in patients with dystrophic EB are common and can produce considerable difficulties in airway management. Equipment and techniques routinely used in the induction or intubation can be the source of hazardous complications in patients with EB besides limitation of mouth opening secondary to scar and contracture [7,13]. Some authors reported that intubation might predispose the patient to high risk of pharyngeal and tracheal lesions [5,14], which result in the formation of intraoral bullae, airway obstruction and extensive hemorrhage. But, if the procedure is performed gently by using a smaller lubricated laryngoscope, the possibility of new bullae formation would be significantly reduced [14,15]. In our case, we performed intubation using the C-MAC® video laryngoscope that is helpful for providing better visualization in patients with anticipated difficult airway. Pediatric patients, as those with difficult airway, can benefit from non-traumatic, rapid, and single-attempt intubation to prevent serious airway complications. Furtunately, we successfully performed tracheal intubation with C-MAC® video laryngoscope on the first trial without any other airway complications. We consider that using the C-MAC® video laryngoscope was helpful for our tracheal intubation and successful airway management, and this factor was the unique aspect of our case in contrast to other pediatric patients with EB.

Monitoring during anesthesia for patients with EB is important and somewhat challenging. In general, the anesthesiologist should pay attention to minimize trauma to the skin and the mucous membranes in handling the patients because of the fragility and vulnerability of skin. Minimal monitoring is recommended for the patients with EB [16]. Electrocardiogram monitoring should be secured with bandages and the adhesive portion should be removed. Pulse oximetry is usually preferred. In some short duration operations, noninvasive blood pressure monitoring (NIBP) can be used with pressure and duration limitation because direct pressure to the skin is not as damaging as frictional or shearing forces. We used NIBP only for the initial blood pressure monitoring and continuous arterial line monitoring during the operation due to skin lesions on the patient's arm. Patients with ocular involvement, on the other hand, are prone to ophthalmic complications such as corneal erosion, corneal blister, corneal laceration and ectropion. Therefore, delicate management is required, including application of moisturizing ophthalmic ointment and moistened protective gauze [15].

Adequate analgesia is also important to prevent excessive movement and additional skin trauma. A multimodal approach using opioids and nonsteroidal analgesics is the most convenient method [17]. Regional anesthesia can be considered in some cases and local infiltration is avoided because of blister formation.

Conclusion

In conclusion, careful anesthetic management is required in patients with EB. Avoiding mechanical injury to the skin and mucous membranes is essential, and delicate preparation and anesthetic planning is needed considering airway comorbidity.

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