The Perioperative Management of a Patient with Epidermolysis Bullosa

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INTRODUCTION

Epidermolysis bullosa (EB) is a rare inherited skin disease, whose chief manifestation is a recurring pattern of skin and mucosal bullous lesions with subsequent scarring after benign amounts of pressure, heat and trauma (1,2). This unique constellation of symptoms presents a challenge to the anesthesia practitioner in the delivery of perioperative care. We present the case of a patient with severe dystrophic EB presenting for the surgical extraction of cataractous teeth.

CASE PRESENTATION

A 22-year-old male, with a past medical history of autosomal recessive dystrophic epidermolysis bullosa (EB), presented to the oral and maxillofacial clinic on referral from a pediatric dentist after an initial chief complaint of pain in multiple teeth. Intraoral and radiographic examinations revealed gross generalized dental caries on the second mandibular molars bilaterally (teeth #18 and #31). The surgical plan included the extraction of these teeth with the possible extraction of the wisdom teeth and bilateral second maxillary molars for hygienic reasons.

Preoperative Anesthetic Evaluation

A thorough evaluation of the patient revealed good exercise capacity with an otherwise negative medical history, except for dystrophic EB. However, he had an extensive surgical history related to EB sequelae. This history included multiple procedures of both the upper and lower extremities secondary to the recurring cycle of blistering, scarring and contracture formation. The physical exam revealed scarring and several amputated digits on his hands and feet. The left hand was remarkable for pseudoarthrodely, with a single digit composed of a fusion of the remaining digits. There were no remaining digits on the right hand (Figure #1). The lower extremity was notable for blistering with no open wounds. The patient had a normal cardiac and respiratory exam. There were no remaining digits on the right hand (Figure #1). The lower extremity was notable for blistering with no open wounds. The patient had a normal cardiac and respiratory exam. 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Anesthetic and Postoperative Management

The anesthetic plan included the use of standard ASA monitors. All the patient’s extremities were padded and wrapped with Coban® (3M, St. Paul, MN) and a blood pressure cuff was placed on the upper arm over a protective layer of NitroVa™ (Coviden, Mansfield, MA). A pulse oximeter was placed on the patient’s earlobe. EKG monitoring was deferred secondary to the patient’s negative cardiac history, good functional capacity, and a pulsatile oximeter waveform.

A smooth mask induction was accomplished with nitrous oxide, oxygen and sevoflurane, using the technique of fiberoptic intubation. Endotracheal intubation was achieved with the addition of midazolam (2 mg), and ketamine (1mg/kg). Anesthesia was maintained with end tidal carbon dioxide monitoring was placed.

An epidermolysis bullosa patient with poor dentition and a limited oral opening due to scarring (Figure #2). Despite poor oral opening, the patient had a Mallampati 1 classification.

DISCUSSION

Epidermolysis bullosa is a heterogeneous spectrum of inherited connective tissue disorders whose incidence is approximately 1 in 50,000 (4). The major subtypes of epidermolysis bullosa include dystrophic, junctional, and simplex EB; of the three, the dystrophic form is the most severe (1). The classic phenotypic findings of skin and mucosal membrane blisters are caused by a mutation in the COL7A1 gene which disrupts the formation of type VII collagen, an adhesion molecule between the basement membrane and the dermis and epidermis (5). Because of the unique manifestation of this disease, this condition presents special challenges in the development of an anesthesia plan.

While there is no standardized treatment for EB, supportive care and prevention have been emphasized to reduce the incidence of skin trauma and thereby reduce associated morbidity (3). Special consideration is required for monitoring due to the extreme fragility of the skin. Tape and adhesives should be avoided if possible. In severe cases, EKG monitoring has been omitted and the pulse oximeter used as its proxy for heart rate sampling (5). When EKG monitoring is used, the adhesive surfaces should be removed and the electrodes should be held on the skin surface using a soft pad (5).

A thorough preoperative airway exam is critical for EB patients as limited oral opening is found in varying degrees in many patients due to previous scarring and subsequent contracture formation. Instrumentation of the airway, as in the present case, should be avoided to reduce the risk of further oral trauma and bulla formation to the tongue or other soft tissues of the oral mucosa. Therefore, when intubation is necessary, nasotracheal fiberoptic intubation has been suggested to be superior over direct laryngoscopy (5). Nasal intubation may also be considered superior over oral intubation because the nasal mucosa is composed of a higher proportion of pseudostratified ciliated epithelium and stratified columnar epithelium, both of which are less prone to bulla formation than the stratified squamous keratinized and nonkeratinized epithelium of the oral mucosa (3).

Finally, depending on the degree of airway trauma, postoperative observation is important. It must be remembered that patients with intraoral pharyngeal lesions from airway instrumentation or intraoperative events are vulnerable to swelling and possible airway compromise. Therefore, prior to discharge, it must be verified that the patients are able to maintain airway patency in the critical initial postoperative hours.

REFERENCES