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Case Report

Pitfalls during Anesthetic Management of Dystrophic Epidermolysis Bullosa; A Case Study

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Abstract

Introduction: Epidermolysis bullosa is a group of genetic diseases that leads to a defect in the dermo-epidermal junction. It affects the skin and mucosa leading to blistering and bullae formation on handling. Similar problems can occur in the mouth and pharynx and this can result in poor mouth opening or tracheal stenosis [1].

Case Report: This case study describes the management and anesthetic plan of a dystrophic epidermolysis bullosa (DEB) case who reached the age of 30 years.

This patient presented with dysphagia to solid food and was posted for dilatation of esophageal stricture under General Anesthesia GA. The patient had a history of difficult intubation, Airway assessment revealed Mallampati class 4, missing teeth, and poor oral hygiene with a thyromental distance of less than 3 cm and restricted mouth opening. The procedure was done with awake intubation followed by GA. Gastroenterologist failed to find an esophageal opening after many trials he abandoned the procedure and advised to do Gastrostostomy.

The patient was shifted intubated to ICU. After 72 hours he was sent back to Operating theatre for extubation with backup surgery team for tracheostomy if needed.

Extubation was uneventful, the patient's family decided to get a second opinion regarding the gastrostomy.

Conclusion: Introducing the best care to DEB patients necessitate Proper planing for each step, and multidisciplinary teamwork, even in simple procedures. Through this study, we will highlight the pitfalls that should be considered in such cases in the future.

Keywords: *Dystrophic Epidermolysis Bullosa; Anesthetic Management; Pitfalls*

Introduction

Epidermolysis is a rare skin disorder that can be inherited or acquired. Patients with heritable forms have abnormalities that cause defects in the anchoring systems of skin layers. The acquired forms are autoimmune disorders in which autoantibodies are produced that destroy the basement membrane of the skin and mucosa. The result is the loss or absence of normal intercellular bridges and separation of skin layers. The separation of the skin layers results in intradermal fluid accumulation and bullae formation [2].

Epidermolysis bullosa can be categorized as simplex, junctional, and dystrophic. In the simplex type, epidermal cells are fragile and mutations of genes encoding keratin intermediate filament proteins underlie the fragility. In the dystrophic types (incidence approximately one in every 300,000 births), the genetic mutation appears to be in the gene encoding the type of collagen that is the major component of anchoring fibrils [3].

The simplex form of epidermolysis bullosa is characterized by a benign course and normal development. By contrast, patients with the junctional form of epidermolysis bullosa rarely survive beyond early childhood. Most die of sepsis. Features that distinguish junctional epidermolysis bullosa from other forms are generalized blistering beginning at birth, absence of scar formation, and generalized mucosal involvement (gastrointestinal, genitourinary, and respiratory). Epidermolysis bullosa dystrophica includes severe scarring with the fusion of the digits (pseudosyndactyly), constriction of the oral aperture (microstomia) and esophageal stricture. The teeth are often dysplastic. Malnutrition, anemia, electrolyte derangements, and hypoalbuminemia are common, most likely reflecting chronic infection, debilitation, and renal dysfunction. Survival beyond the second decade is unusual [3].

Case Report

A case 30 years old male, 37 kg height is 160 cm and BMI of 14.7 kg/m^2, diagnosed as DEB, he had a strong family history of the same disease (his cousin).

Significant preoperative findings were generalized scars, foot ulcers, Mitten hand deformity, Limited mouth opening and neck extension, dystrophic teeth, Skin ulcers in his back, feet, and legs. This patient required frequent hospital admissions for anemia, submandibular infection, and multiple dental extractions.

He had previously undergone different procedure under general anesthesia and sedation.

The patient had survived from a previous cardiac arrest in the recovery room after a dental extraction under GA.

His previous esophago-gastro-duodenoscopy OGD revealed stenosis in the upper esophagus, Barium swallow showed upper esophagus stricture and aspiration from supra-glottic level.

Recently the patient noted to have shortness of breath with exertion, coughing while eating, he was evaluated by CT chest and neck which revealed: 1- Mild abnormal mucosal thickening and enhancement involving aryepiglottic folds extending to involve the pharyngo-esophageal junction and post cricoid region of hypo-pharynx with a suspected significant stenosis involving the cervical esophageal inlet. 2- Large calculus in the left mandibular gland. 3- Ground glass opacities were noted involving the right middle lobe suggestive of an acute process (aspiration). Chest team advised to start antibiotics for one week as well as bronchodilators and corticosteroids.

Aspiration from ruptured blisters is common findings like in our patient who had wheezy chest, frequent cough but he was maintaining normal spo_2 on room air.

The patient and the family were interviewed before the procedure they understood the difficulty we might face with the airway and agreed upon our plan; which was awake intubation or abandoning the procedure in case of failure to intubate, with the surgical airway as a backup in case of life-threatening.

The patient transferred himself to OT table, Anesthesia was started with basic monitoring ECG was done using the gel non-adhesive part of ECG leads (Figure 1) and fixed by gauze padding and crepe bandage around the chest while the patient is sitting (Figure 2), thick layer of cotton was used below the non-invasive cough (Figure 3) and a pulse oximeter was attached to his ear lobule , iv access was secured using an non-adhesive crepe bandage, all pressure areas were padded with cotton to avoid any friction or pressure to the skin.

All difficult airway equipments were available, special wet gauze, silver sulphadiazine cream and Vaseline gauze for face mask (Figure 4).

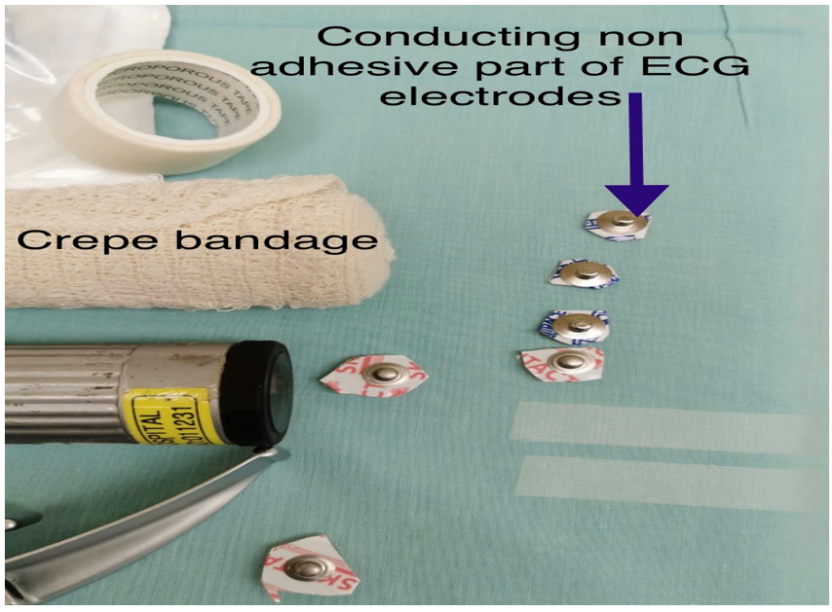

Figure 1: *Non-adhesive conducting parts of ECG electrodes.*

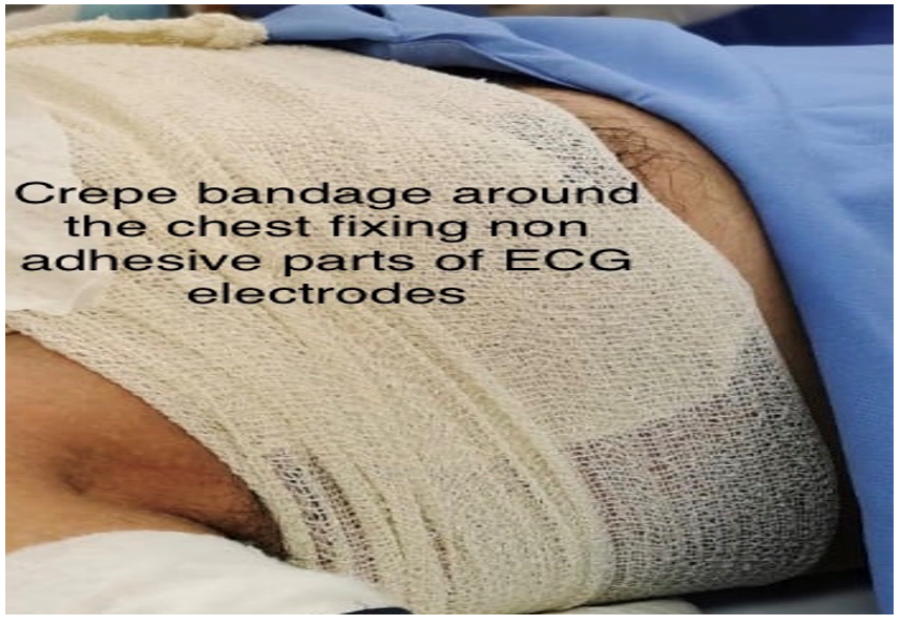

Figure 2: *ECG electrodes fixation in DEB.*

Figure 3: *Cotton padding under non invasive blood pressure cuff.*

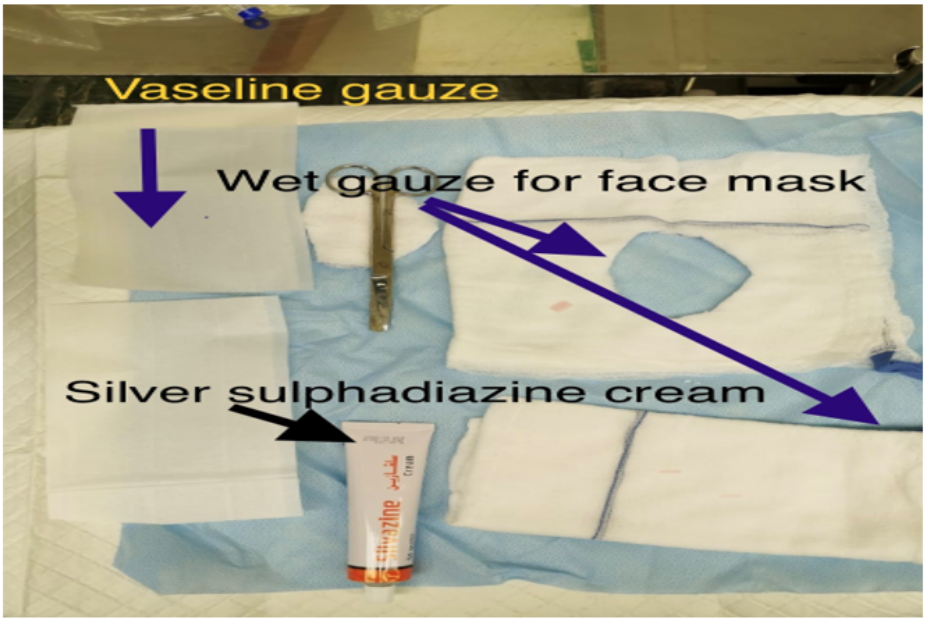

Figure 4: *Tools helping in airway in DEB.*

Preoxygenation and lidocaine nebulization were done before start.

Using the spray as you go technique, glycopyrrolate, dexamethasone, and 10 mg of ketamine, fibro optic intubation was done, Intubation was difficult; distortion of airway anatomy noted and the epiglottis was fibrous abnormally positioned as if guarding vocal cords. The patient was intubated with 6.5 ETT and was secured with non-adhesive lubricated bandage Propofol and cisatracurium were given immediately after intubation.

The procedure started and The gastroenterology tried to find the esophageal opening but failed to find even evidence or track to follow, and then he gave up and advised to do gastrostomy in another sitting.

The patient kept Vitaly stable during and after the procedure, Then he was shifted ventilated intubated to ICU as planned.

The intensive care team and gastroenterologist talked to the family regarding the options of elective gastrostomy and tracheostomy as the last treatment options, but the family didn't agree and they decided to take a second opinion and asked for discharge after extubation.

At that time there was the issue of emergency tracheotomy if needed during extubation was raised from the ICU side, in spite of the initial agreement of the family, then they refused to give consent. Two ICU consultants and administration and legal affairs representatives and the surgeons all attended a meeting with the family after that they agreed to give the consent for emergency tracheostomy only in life-threatening conditions, patient was transferred to OT after 72 hours for extubation with all difficult airway facilities, two senior anesthesiologists, and in the presence of surgeons and emergency tracheostomy if needed.

Extubation was uneventful. nebulization and corticosteroids were used in recovery.

Discussion

DEB is a genetic disorder which results in skin damage after minor trauma patients with this condition usually suffer from blistering in mucosal membranes, especially in the esophagus, repeated trauma and blistering then ulcers and healing causes strictures in the upper airway, esophagus and urethra as well.

Preoperative evaluation should focus on airway assessment, cardiopulmonary and patient's present condition; blistering, nutritional status, infection, and the use of steroids.

Regarding the care of skin; Avoiding mechanical injury to the skin and mucous membranes are essential in anesthetic management. Shearing forces applied to the skin result in bullae formation, while compressive forces to the skin are tolerated [4]. Bed sheets on which patients are placed should be crease-free.

Generous padding of the pressure points should be done with soft cotton, the perioperative blister should be treated with liquid paraffin, silver sulphadiazine, or steroids. In the ambulatory setting, small procedures can be performed under analgesia and sedation without airway manipulations [5].

Eye protection is important as EB patients are prone to ocular manifestations like scarring of the eyelids, After application of a moisturizing ophthalmic gel, eyes should be covered with moistened gauze to protect from mechanical trauma [6].

Any airway instruments need to be lubricated with water-based jelly, oropharyngeal airways should be avoided if possible, face masks should be lubricated.

Endotracheal tubes should be smaller than expected, softened and well lubricated. Vigorous suctioning of the oral cavity is not recommended [7].

Regional anesthesia is a good alternative whenever possible to avoid airway manipulation, care should be taken during cleaning the puncture site; the preparatory solution is to be poured over the area [8]. Struggling during waking up from anesthesia can lead to blister formation, Pain relief should be adequate so to prevent excessive movement or irritability [9].

Oxygen mask can be avoided or applied over Vaseline gauze as the sharp edges of the face mask can cause skin injury. This Patient also had generalized scars, lost toes and fingers with ulcers on the skin of his feet that required the patient to transfer himself to OT table and to be awake during the padding for his body ulcers.

In DEB patients nasal, oral, pharyngeal, and tracheal manipulations should be kept to a minimum to avoid airway damage.

In this patient, he had a history of cardiac arrest in the recovery post-dental procedure. We couldn't find any comments on this incident in the previous records.

When we take the history, the patient suggested that he might have been exposed to trauma which resulted in blister after surgery and this could be large enough to suffocate him, these were the patient own words commenting on the previous incident.

The initial Anesthesia plan based on patient history, airway assessment and the urgency of the procedure was to do awake fibro-optic intubation and to minimize upper airway manipulations, and In case of not being able to intubate the patient, we will cancel the procedure, on the other hand, if we succeeded in intubating the patient we will shift him intubated to ICU for 48 hours the expected time to the blisters to disappear.

This was a safe plan for dealing with expected difficult intubation and to care for this patient after trauma caused by intubation and multiple trials of upper GI scope.

But the plan was missing the possibility of tracheostomy during extubation, which was raised immediately by the ICU team on admission, Preparing for surgical airway during extubation, family counseling and consents were not part of the plan which required later more efforts and time to prepare.

Also, there was no multidisciplinary plan to deal with what happened in this patient; like in case of inability to access the esophagus what will be the next step and why preparations and consents were not ready for doing gastrostomy instead of exposing the patient to that risk of intubation and extubation.

Despite all the potential complications with anesthesia for patients with epidermolysis bullosa intraoperative management is associated with surprisingly few complications. this is especially true when care is provided at a center experienced with the management of patients with epidermolysis bullosa [2].

Conclusion

In conclusion; proper perioperative multi-disciplinary planning and communication among healthcare teams extremely required in the anesthetic management of DEB to achieve the best patient outcome.

Source Support

Nil.

Conflict of Interest

There are no conflicts of interest.

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