Epidermolysis Bullosa Dystrophica and Anesthesia

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Abstract

We report the use of ketamine anaesthesia in a 41 years old male patient with epidermolysis bullosa dystrophica (EBD) scheduled for inguinal lymphadenectomy.He had EBD diagnosis for 25 years, with a history of laryngeal, tracheal and oesophageal bullae. On physical examination he had numerous blisters in dorsal and lumbar region.According to the literature, general anaesthesia is preferable in most of the EBD patients with low incidence of complications. Regional anaesthesia is an option with the recommended precautions, but it is limited to patients without lesions in punction areas.Ketamine seemed a good option, because of the nature of the intervention as it obviates the need of airway manipulation.This patient has overtaken the expected longevity of the disease and so we had the obligation of continuing the skin care done by him and his dermatologists, and so, he represented a huge challenge for us.

CASE REPORT

Epidermolysis bullosa dystrophica is a rare hereditary disease characterized by the formation of mucocutaneous blisters after light or nonexistent trauma. The estimate incidence is 1:300000 births per year. The disease affects mostly the stratified squamous epithelium, although any mucous membrane can be affected [1,2]. Some patients have generalized blisters on skin and mucous membranes resulting in atrophic scars, conditioning important morbidity and mortality. There are multiple surgical and medical problems associated, being the anesthetic risk attributed mainly to the oropharingeal involvement, airway management, patient mobilization and surgical positioning [3]. There is no cure for this disease. The treatment is supportive and the main objective is to avoid the skin and mucous membranes trauma and to compensate the associated multiple organic imbalances [4]. The surgical treatment is important in the prevention of debilitating sequelae or in the resolution of associated complications, although with considerable anesthetic risks. The malignant degeneration of the stratified squamous epithelium in the adulthood is frequent and the main cause of death in late phases of the disease.

A 41 year olf male, Caucasian, 45 Kg weight and 1.60 m height, proposed for surgical resection of left inguinal adenopathy. Diagnosis of neonatal autosomal recessive EBD for 25 years with follow-up on Dermatology and multiple hospital admissions. Last hospital admission motivated by painful adenopathy on the left inguinal region with two months of evolution that was biopsed and revealed squamous cell carcinoma (SCC).

In the preanesthetic evaluation, the patient presented conscious, calm and cooperative with normal vital signs and cardiopulmonary auscultation.

Sindactilia of the hands and feet (figure 1 and 2).

Figure 1

Figure 1. Syndactyly of the hands

Figure 2

Figure 2. Syndactyly of the feet

Ulcerated, blistered and hemorrhagic wounds of the lower and upper members, dorsal and lumbar region (figure 3), not affecting the head and neck.

Airway: microstomia, Mallampati grade II, cervical and jaw mobility maintained.

Anesthetic-surgical history of maxillofacial surgery for dental implants with local anesthesia. Two surgical resections of SCC in the left knee under local anesthesia. History of blister formation in the larynx, trachea and oesophagus.

Family history: no consanguinity, first line cousin in the maternal lineage with EBD.

Analytics: hemoglobin of 10.8 mg/dL; chest X-ray and electrocardiogram without alterations..

The patient was classified ASA III. Preanesthetic medication: lorazepam 1,0 mg orally on the day before and on the morning of surgery.

On the arrival to the theatre the patient was under medical supervision since the transfer zone to the operative room, cooperative, level II of Ramsay sedation scale and was encourage to active collaboration in position changes. Monitorization: cardiac electrodes without adherent surface, pulse oximetry with ear probe and non invasive blood pressure cuff with underlying skin wrapped in gauze with manual evaluation every 10 minutes. Peripheral intravenous access on the left forearm fixed in place with silk and wrapped in gauze.

Anesthetic technique: induction with intravenous ketamine in fractionated doses (125 mg total) associated with midazolam 4 mg. Intravenous administration of atropina 0.5 mg for antisialogogue effect and acetaminophen 1 g for post-operative analgesia.

On the intraoperative period the patient was hemodinamically stable and in spontaneous ventilation with O_2 atmosphere. After the end of the surgical procedure the patient was transferred to the recovery room and monitorized with pulse oximetry, maintaining a darkened and calm environment. Postoperative analgesia with acetaminophen 1 g and tramadol 100 mg intravenously. He had no pain or other complaints.

In the first postoperative day the patient referred amnesia for the perioperative period, light left inguinal pain and auditive and visual hallucinations during the immediate postoperative period. There was no psychomotor agitation. He had no

blisters related to the intervention including in the local of electrodes and non-invasive arterial pressure cuff placement or related with position changes.

The goal of the anesthetic approach in these patients is the "no touch principle". In the preanesthetic evaluation a clinical history should be carried out and a complete physical exam with evaluation of the disease and its general consequences. Blistered wounds presented before surgery should be registered for identification of eventual iatrogenic wounds. The evaluation of the airway is crucial because in the majority of the cases there is a difficult airway conditioned by the scars on the face. The patient previous anesthetic procedures, if they exist, should be reviewed, for documentation of previously difficult airway. The adults should come for the operative room awake and help in their positioning. In the intra-operative period the protection of all pressure points in the skin is essential. All the adhesive material must be banished. All the materials in contact with the patient should be well lubricated. Protection of the eyes with an ophthalmic ointment and a vaselinated gauze is essential. The monitorization 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. In our case we choose the pulse oximetry (ear probe), non invasive blood pressure and electrocardiogram. For monitorization of the non invasive blood pressure it's necessary to place several soft non adherent gauzes under the cuff. The adhesive of the electrodes must be removed.

The intravenous catheter should be fixed with nastro, vaselinated gauze or line of suture.

As regards to the anesthetic technique, general anesthesia is the technique of choice in the majority of the studies published [1,3,4,5], with a low incidence of airway blisters formation. The recent revision articles accentuate the security of the endotracheal intubation, since the relative precautions to lubrication, smooth manipulation and fixation of the materials are taken. The fiberoptic and tracheotomy equipment should be prepared in the possibility of failed attempts of endotracheal intubation. During the extubation, the aspiration of the oropharinge is contraindicated by the risk of big and hemorrhagic blisters formation.

The regional anesthesia is a valid alternative to the general anesthesia because there is no need of airway manipulation [1,3,4,5]. These techniques require the same principles of care as general anesthesia in their execution and positioning of patients, with correct inspection of puncture and compression sites. The presence of blisters and signs of infection at sites of puncture contraindicates these procedures; this was the motive why we didn't choose a regional technique in this patient. The subcutaneous infiltration with a local anesthetic is contraindicated due to the possibility of epidermis dissection and formation of big blisters. After puncture, the area should be covered with fat gauze. The superficial nature of the surgical procedure, the contraindication to regional anesthesia and the history of oropharingeal mucous membrane involvement lead us to prefer ketamine anesthesia in this case. Although there are limitations to its utilization, namely the psychomotor agitation and psychiatric alterations in the emergency of anesthesia, with increased risk of trauma, this can be avoided with association of a benzodiazepine.

In the postoperative period the utilization of oxygen rich atmosphere is preferential to the utilization of oxygen facial masks and nasal probes. The vigilance should settle in the risk of airway obstruction due to a big blister or hemorrhage formation. The analgesia cannot be forgotten as pain causes agitation with risk of iatrogenic lesions.

CONCLUSION

This patient represented a big challenge for everybody who contacted with him in the theatre since he had already surpassed the expected longevity of the disease, being our obligation to protect him against new iatrogenic wounds and guaranteeing the continuity of the skin care. It is fundamental to have an integrated and multidisciplinary perspective of this disease as we tried to do in the case presented here.

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