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Combination of Ketamine with Dexmedetomidine Infusion as Non-operating Room Anesthesia/Opioid-Free Anaesthesia for Oesophagial Dilatation in a Paediatric Patient of Epidermolysis Bullosa Dystrophica: A Case Report

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Abstract

Introduction: Dystrophic Epidermolysis Bullosa (DEB) poses major challenges, especially for anesthetic management. The main challenge faced is a pediatric patient with a difficult airway requiring general anesthesia. Any airway manipulation leads to immediate and significant trauma with bullae formation, thereby necessitating the need for postoperative ventilation and prolonged hospital stay. It is advisable to avoid any airway interventions, if possible.

Case Presentation: An 11-year-old male child (weighing 15 kg) presented with non-bilious vomiting for 3 - 4 days and dysphagia for solids for 2 - 3 months. Personal history revealed the diagnosis to be a case of DEB at the age of 10 years (genetic testing) with persistent iron deficiency anemia. The patient underwent esophageal dilatation 6 months ago under general anesthesia (propofol, fentanyl on spontaneous respiration), requiring blood transfusion and an intensive care unit (ICU) stay for a day after the procedure. Dexmedetomidine (Dex) injection was started as a low-dose infusion (0.2 μg/kg) 10 minutes before the start of the procedure, followed by (0.2 μg/kg/hour). Before upper gastrointestinal (GI) scope insertion, ketamine injection (0.66 mg/kg) 10 mg intravenous (IV) bolus was given. Upper GI endoscopy and esophageal dilatation were performed in half an hour without any complications. Dexmedetomidine infusion was stopped after the removal of the scope, and the patient awakened within 3 - 4 minutes. He remained hemodynamically stable throughout the entire procedure. Recovery was uneventful, and the patient was obeying commands with no signs of respiratory depression, postoperative nausea/vomiting (PONV), or pain. Vital signs remained stable in the post-anesthesia care unit (PACU), and then the patient was moved to the ward.

Conclusions: To date, there is no reported case of DEB being successfully managed with only two drugs, dexmedetomidine infusion and a single dose of ketamine for maintaining spontaneous ventilation (opioid-free anesthesia/OFA) with no airway intervention/manipulation, especially in non-operating room anesthesia (NORA) setting.

Keywords: Epidermolysis Bullosa Dystrophica (EBD), Ketamine, Dexmedetomidine, NORA/OFA

1. Introduction

Epidermolysis Bullosa (EB) was first described in 1870 by Ferdinand Ritter von Hebra, an Austrian physician/dermatologist and also the discoverer of the causative organism of scabies (1). A loss in function of keratin, collagen, and laminin in the skin leads to inherent weakness, fragility, and dehiscence at the dermal layer at which they are prevalent. In addition, the skin becomes a less effective functional barrier to infection, and there is impaired healing.

Notwithstanding the multiple different genetic types of EB, three major histological groups have been described: Dystrophic EB (EBD), EB simplex (EBS), which is the most common, and junctional EB (JEB) (2). These groups are based on the precise ultrastructural level of the dermis at which the weakness and, therefore, dehiscence occurs.

The EBD subtype is characterized by a tendency for bullae formation, and the bullae tend to heal, forming significant amounts of scar tissue. This often leads to the fusion of digits and significant amounts of cutaneous

scar tissue; however, in addition, since collagen VII is found in many non-cutaneous membranes, oesophagial joint and peri-orbital contractures are observed. As a result, DEB is the most common subtype presenting for surgical treatment (3). Herein, we present one such case requiring frequent non-operating room anesthesia (NORA) management under opioid-free anesthesia/opioid-sparing strategy (OFA/OSS) (4).

2. Case Presentation

An 11-year-old male child (weighing 15 kg) presented with non-bilious vomiting for 3 - 4 days and dysphagia for solids for 2 - 3 months. He was the third issue of a non-consanguineous marriage, full term with normal vaginal delivery, weighing 2.5 kg, with a neonatal intensive care unit (NICU) stay for 2 days for extensive skin lesions.

Personal history revealed the diagnosis to be a case of DEB at the age of 10 years (genetic testing) with persistent iron deficiency anemia. Family history revealed that the oldest sibling, with confirmed DEB, died at 2 months of age. The second sibling is normal with no features of DEB.

The case underwent oesophageal dilatation 6 months ago under general anesthesia (propofol, fentanyl on spontaneous respiration), requiring blood transfusion and an intensive care unit (ICU) stay for a day after the procedure.

General examination showed tachycardia (120-130/min), afebrile, pallor, chronically malnourished with severe stunting and wasting (age and weight below 3rd centile), multiple crusted plaques (hyper and hypopigmented with active oozing), all over the body. There were multiple skin bullae and exposed subcutaneous tissue with blisters over the entire face, circumferentially covering the neck, both hands/legs, abdomen, and back.

Airway examination showed a mouth opening of 1 finger with the inability to protrude the tongue and Mallampati grade IV. Apart from iron deficiency anemia and hemoglobin of 9.4 gm/dl, other routine investigations were normal. Upper gastrointestinal (GI) endoscopy conducted previously showed mid-oesophagial stricture. He was on tablet losartan and tablet levocetirizine. The patient was given fitness under the American Society of Anesthesiologists (ASA) grading III E. The case was planned for oesophageal dilatation under deep sedation/GA after relevant pediatric and gastroenterologists' opinions.

On the day of the procedure, intravenous access was secured on the right hand with gauze pieces immersed in Vaseline and bandages. After taking written informed consent, the patient was moved to the procedure room carefully with gentle handling.

Monitors, electrocardiogram (ECG)-lubricated central gel part placed on healthy skin, oxygen saturation (SpO_2), and NIBP cuff (padded with cotton) were attached on appropriate skin sites.

The plan was to maintain anesthesia on spontaneous ventilation and to avoid airway manipulation. Dexmedetomidine (Dex) injection was started as a low-dose infusion (0.2 µg/kg) 10 minutes before the start of the procedure, followed by (0.2 µg/kg/hour) and O_2 started with nasal prongs at 2 lit/min. The heart rate stabilized between 90 and 100 per minute after 10 minutes and remained so throughout the procedure.

Before upper GI scope insertion, a ketamine (0.66 mg/kg) injection of 10 mg intravenous (IV) bolus was given. Upper GI endoscopy and oesophageal dilatation were performed in half an hour without any complications.

Dexmedetomidine infusion was stopped after the removal of the scope, and the patient awakened within 3 - 4 minutes. He remained hemodynamically stable throughout the entire procedure.

Recovery was uneventful, and the patient was obeying commands with no signs of respiratory depression, postoperative nausea/vomiting (PONV), or pain. Vital signs remained stable in the post-anesthesia care unit (PACU), and then the patient was moved to the ward.

3. Discussion

Since DEB child presents numerous anesthetic challenges, various modalities have been used based on available resources, the need for airway manipulation, and surgical procedures. The main concerns are to avoid trauma to fragile skin/mucus membranes, right from transportation to attaching monitors, and gentle handling of the airway to avoid bullae formation.

Non-operating room anesthesia poses major challenges, such as unfamiliarity of venue, paucity of equipment/human resources, and minimum intervention (4). Opioid-free anesthesia is a desirable strategy with obvious advantages, especially employing the dexmedetomidine/ketamine (dex-ket) combination.

The mechanism of action of Dex is unique and differs from those of currently used sedatives, including clonidine. Pre-synaptic activation of α2 receptors in the brain and spinal cord inhibits the release of norepinephrine and inhibits neuronal firing, terminating the propagation of pain signals and

producing analgesia and sedation. The major problem is hypotension and bradycardia. (5, 6). Our aim was to start a continuous infusion of dexmedetomidine at a minimal rate before induction to minimize the side effects of bradycardia, hypotension, and postoperative sedation (7).

The addition of ketamine (N-methyl-d-aspartate [NMDA] antagonist) helps with its effects of hypnosis, followed by increasing sedation and unconsciousness at higher doses, intense analgesia (or more accurately, anti-nociception), increased sympathetic activity to nullify Dex actions, and maintenance of airway tone and respiration (8). A combination of just these two drugs helped achieve the desired level of deep sedation required for the procedure, always maintaining hemodynamic stability and spontaneous respiration without airway manipulation, along with adequate analgesia and faster recovery.

In conclusion, despite all the potential challenges of DEB/NORA, innovative intra-procedural management is associated with fewer adverse events. We propose using a low-dose dexmedetomidine infusion (0.2 μg/kg) for 10 minutes, followed by (0.2 μg/kg/hour), along with ketamine injection as OFA/OSS to avoid airway manipulation and smooth intra-operative/procedural course with immediate and clear-headed recovery and added benefit of excellent analgesia.

Footnotes

Authors' Contribution: MMP conceived and designed the evaluation and drafted the manuscript. AG conducted the case under the supervision of MMP and helped draft the manuscript. MMP conducted most of the literature search and finalized the manuscript. AG revised the manuscript. MMP finalized the manuscript. All the authors read and approved the final manuscript.

Conflict of Interests: Authors declared no conflicts.

Data Availability: The data presented in this study are uploaded during submission as a supplementary file and are openly available for readers upon request.

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Informed Consent: This was not a study but a simple case report. Consent for the conduct of anesthesia was obtained.

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