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Case Report

Anesthesia Management Of A Patient With Hereditary Angioedema Under Dexmedetomidine: A Case Report

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ABSTRACT

Anesthesia management of a patient with hereditary angioedema under Dexmedetomidine: A Case Report.

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Hereditary angioedema (HAE) is a life-threatening rare autosomal dominant (AD) disorder caused by C1-esterase inhibitor (C1-INH) deficiency or dysfunction. HAE is not related to mast cell and histamine pathway; as a result, it does not respond to antihistamines, glucocorticoids, or epinephrine. Attacks may occur spontaneously or after a trauma. The upper airway swelling and laryngeal edema cause asphyxia, which can be life-threatening. Treatment of HAE includes C1-INH concentrate, recombinant human C1-INH, bradykinin B2-receptor antagonists, recombinant plasma kallikrein inhibitors, fresh frozen plasma (FFP) and androgen steroids. In this case we report anesthetic management of a 62 years old female patient with HAE, undergoing endoscopy and colonoscopy, for which highly selective α2 adrenergic receptor agonist dexmedetomidine intravenous (IV) infusion was used. We administered icatibant (bradykinin B2-receptor antagonist) for prophylaxis, and FFP was prepared in case of use. Anesthetic management and preoperative preparation are critical for the patient with HAE. Difficult airway management should be kept in mind in case of airway edema and early intubation.

Keywords: Hereditary angioedema, dexmedetomidine, anesthetic management, icatibant, fresh frozen plasma.

INTRODUCTION

Hereditary angioedema (HAE) is a life threatening rare AD disorder caused by C1-esterase inhibitor (C1-INH) deficiency or dysfunction[1]. The estimated prevalence of HAE is 1:60.000, with ranging from 1:10.000 to 1:150.000, and it has been reported in all sexes and races[2-4]. C1-INH deficiency results in excessive bradykinin production, a potent vasodilatory molecule, and vascular permeability-enhancing effects[5]. HAE is not related to mast cell and histamine pathway; as a result, it does not respond to antihistamines, glucocorticoids, or epinephrine. Attacks may occur spontaneously or after a trauma (endoscopy, intubation, dental procedures). Angiotensin-converting enzyme (ACE) inhibitors and non-steroidal anti-inflammatory drugs (NSAIDs) may trigger HAE attacks as well[6,7]. Clinically it most often affects the skin (without pruritus); the upper airway, which may cause laryngeal edema, asphyxia and sudden death; the gastrointestinal tract bowel wall edema causes abdominal pain, vomiting, nausea, and diarrhea.

HAE is often self-limited and resolves in two to five days without treatment but it can be life-threatening as well. The first-line therapies are C1-INH concentrate or recombinant human C1-INH. In addition, bradykinin B2-receptor antagonists (icatibant) and recombinant plasma kallikrein inhibitors can also be used as first-line therapies[8-12]. FFP can be used at 10-15 mL/kg as a second-line therapy if the first-line therapies are unavailable[13]. Prophylactic androgen steroids decrease attack frequency and severity and can be used for long-term prophlaxis[4].

Dexmedetomidine is a highly selective α_2 adrenergic receptor agonist, and it is frequently used in intensive care units (ICU) for sedation and prevention of delirium and provides conscious sedation and analgesia without respiratory depression[14].

When left undisturbed the patient sleeps in a natural state of sleep, but the patient can be awakened with a slight stimulus.

In this case, we aim to present anesthetic management of a 62 years old female patient with HAE undergoing endoscopy and colonoscopy. Icatibant was administered subcutaneously before the procedure for prophylaxis, and FFP was prepared in case of need. We used highly selective α_2 adrenergic receptor agonist dexmedetomidine IV infusion for sedation. We also discussed a prophylactic approach to HAE management accompanied by literature.

CASE PRESENTATION

A 62 years old female patient (Weight: 74 kg, Height: 158 cm, BMI: 29.64) with a history of hypertension, bilateral mastectomy for breast cancer, primary immune deficiency and HAE, which was diagnosed at the age of 58, was admitted (April 2022) in our institution. She was receiving intravenous immunoglobulin

(IVIG) treatment for primary immune deficiency every 15 days. Additional her drug treatment included candesartan cilexetil, escitalopram, esomeprazole, and icatibant (a bradykinin B2-receptor antagonist) for HAE treatment. In her medical history, she mentioned a spontaneous upper airway attack, ten days prior to a dental surgery one month before admission, and a spontaneous laryngeal edema and vocal cord edema three years before admission, which caused dyspnea and led her to the emergency hospital department. Written and verbal consent was obtained from the patient before recording the information in this report.

Her physical examination was normal. The results of preoperative laboratory examination, electrocardiography and chest radiography were normal. Her blood pressure (BP), peripheral oxygen saturation (SpO_2), body temperature, respiration rate (RR) and heart rate (HR) were in a normal range. Anesthetic management was planned before the endoscopy and the colonoscopy procedure. FFP was prepared in case of use for HAE attack. After careful pre-anaesthetic evaluation and in agreement with the Immunology-allergology department, icatibant was administered 30 mg subcutaneously 10 minutes before the procedure. Standard monitoring was applied (HR, BP, RR, SpO_2), and bispectral index (BIS) monitoring was used to evaluate the patient's level of consciousness. Emergency airway equipment, endotracheal tubes and video laryngoscope, fiberoptic bronchoscope, and tracheostomy set were prepared in case of use. A port catheter was used for administrating medications. Dexmedetomidine 0.5 mcg/kg IV bolus was given 10 minutes before the anesthesia induction. Lidocaine 1 mg/kg IV, midazolam 2 mg IV and propofol 0.5 mg/kg IV were used for the anesthesia induction, and granisetron 2 mg IV was administered 10 minutes before the end of the process. Dexmedetomidine 0.1-0.7 mcg/kg/h IV infusion was administered during the endoscopy and colonoscopy. We tried to keep the BIS score in 60-80 range. The gastroenterologist performed a gastric biopsy and polypectomy from the colon. She received a 500 mL isolyte - S IV solution. The procedure lasted 60 minutes, the patient remained hemodynamically stable, and no complications occurred during and after the endoscopy and colonoscopy. The patient was followed up for 1 hour at Post-Anesthesia Care Unit (PACU), and she was discharged home uneventfully.

DISCUSSION

HAE is a rare, self-limited, life-threatening AD disorder. It may be triggered by trauma, dental work, intubation or spontaneously. The mortality rate is approximately 30% from laryngeal swelling. Laryngeal edema causes asphyxia and sudden death. In our case, she had spontaneous laryngeal edema and vocal cord edema three years ago, which caused dyspnea to go to

the emergency room. Replacing the C1-INH protein is the main treatment point for HAE. Hosokawa R et al. reported no complication developed during sedation with propofol and C1-INH concentrate administration 1 hour before the dental procedure[15]. Recombinant plasma kallikrein inhibitors and androgens can be used for long-term and short-term prophylaxis. Treatment of HAE includes FFP, which is another pre-surgical option. Bradykinin B2-receptor antagonist, icatibant, can be used for long-time and short-time prophylaxis. Bang YS et al. reported total laparoscopic hysterectomy without any airway complication. They treated the patient with danazol before the intubation and transfused three units of FFP during the surgery[16]. We administered 30 mg icatibant subcutaneously and prepared FFP in case of use for HAE attack.

It is crucial to have spontaneous breathing to not interfere with the airway, which may trigger HAE.

Dexmedetomidine was used for the sedation during the endoscopy and colonoscopy, and spontaneous breathing was not suppressed. The patient recovered quickly at the end of the procedure. Dexmedetomidine provides more physiological sedation, and we recommend dexmedetomidine sedation for non-operating room anesthesia (NORA).

Proper medications, a multidisciplinary approach and difficult airway management are essential for successful anesthesia management and patient safety. In HAE recurrent skin swelling without pruritus or upper airway edema, which is unresponsive to antihistamines or epinephrine therapy, should suggest HAE.

CONCLUSION

Anesthetic management and preoperative preparation are critical for a patient with HAE. C1-INH concentrate, icatibant, FFP or other medicines should be readily available for prophylaxis and therapeutic purposes. Difficult airway management should be kept in mind in case of airway edema and early intubation. Drugs that do not suppress spontaneous breathing should be chosen to avoid airway manipulation. Dexmedetomidine offers more physiological sedation, and we recommend dexmedetomidine sedation for NORA.

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Authors' contributions:

TÖ collected paper's data and is the lead author. BE collected paper's data. EZ contributed to planning the paper. TÖ contributed to planning the paper. ÇN drafted the paper, contributed to planning and the critical revision of the paper. All authors read and approved the final manuscript.

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