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Case Report Open Access

Airway Management in a Patient with Infantile Systemic Hyalinosis

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Abstract

Infantile Systemic Hyalinosis (ISH) is a rare inherited disease that is found among some families in Arab region where consguinity is not uncommon. Patients with ISH have distinctive clinical features that label them as difficult intubations. This includes limited mouth opening, restricted neck movement, protrusion of the mandible, and gingival hyperplasia. We describe the anesthetic management in 24 year old lady who presented for an elective procedure and the different ways to approach these patients.

Introduction

Inherited Systemic Hyalinosis (ISH) is a rare systemic disease, but it is more common in the Arab world where consguinity plays a role [1]. ISH is inherited in an autosomal recessive manner [2]. It is characterized by hyaline deposits in the papillary dermis and other tissues. Patients with ISH usually present with distinctive clinical features including the following: hyper pigmented skin over bony prominence, progressive contractures, and severe pain with passive movements, gingival hypertrophy, and skin nodules over the face, neck and perineal region, depressed nasal bridge, ear malformation, coursed facial features, and failure to thrive [2]. From the above-described features, managing the airway in these patients can be quite challenging for anaesthetists. It requires special preparations and the presence of an expert anaesthetist.

Case Report

A 24 year old patient with normal mental capacity, a known case of ISH, diagnosed at the age of 6 months, was scheduled for elective surgery for removal of subcutaneous nodule (5 cm×5 cm) at the occipital region. She is not known to have any other medical problems or allergies. She is not on any medication. She had previous surgery to remove subcutaneous nodule on the right side of the chest wall 2 years ago, but there was no anaesthetic report available. The patient requested general anaesthesia for the procedure.

On examination, the patient was vitally stable. Her BP was 113/82, HR 78, maintained SpO_2 95% on room air. Her weight and height were 21 Kg and 98 cm, consecutively. Examination of the airway showed that there were no visible nodules, but there was limited mouth opening (<2 cm), decrease in thyromental distance (<4cm), a short neck, restricted TMJ and neck movements and narrow nares (Figure 1). She had Mallampati score of IV with gingival hypertrophy, chipped teeth and poor oral hygiene. Other systemic examinations revealed kyphoscoliosis at the back and fixed contractures at all limbs with significant growth retardation and difficult intravenous access. The patient was admitted one night before the surgery. High-risk consent was obtained regarding the possibility of difficult airway. Premedication included Ranitidine 25 mg IV, Metoclopramide 3 mg IV, and Midazolam 0.5 ml (5 mg) syrup PO.

Intraoperative plan was to maintain spontaneous respiration with inhalational anesthetics and to use Laryngeal mask airway (LMA), since the procedure was short (<30 mins) and the anticipation of difficult intubation. After connecting the patient to the monitors, we managed to insert 22Gauge IV cannula after multiple trials. Patient was given 1 mg IV midazolam followed by induction with sevoflurane and oxygen. There was difficulty in mask ventilation. A supplement of 50 mg IV Propofol was given before trying to insert size 3 LMA. There was some difficulty in inserting the LMA so we decided to replace it with size 2.5 LMA. There was the obvious leakage sound heard around the tube and $etCO_2$ trace on the monitor was not found. The patient started to desaturate SpO_2 progressed rapidly down to 44. The appropriate action that was taken was to awake the patient and abort the procedure. The patient was rescheduled for the next day to do awake fiberoptic intubation. The plan was to prepare the patient properly by giving her Hydrocortisone 25 mg IV to minimize airway edema, Glycopyrulate 200 Mcg IV to minimize the secretions.

The next day for preparation for the awake fiber-optic, the patients' airway was anesthetized using nebulized 5 ml of 2% of Lidocaine, 20 minutes before the procedure. Intraoperative, 10 mg IV Ketamine was given for mild sedation. We used adult size fiber-optic scope connected to video monitor and the fiber-optic was preloaded with size 6.5 ETT. The operator was able to visualize the vocal cords; multiple nodules were noticed along the airway tract. The fiber-optic passed successfully between the vocal cords and the carina was visualized. The ETT did not pass through the vocal cord despite rotating the ETT medially and pushing down on the tongue with a tongue depressor. We decided to try with a smaller ETT. A second attempt was done using an eyepiece pediatric fibro-optic scope preloaded with a tube size 5.5. Again the operator was able to visualize the vocal cords. The flexible tip of the fiber-optic would get stuck between the nodules located in front of the arytenoids. Negotiating the fiber-optic to pass this obstacle was not possible because of the twisting and flexing of the fiber-optic would make it impossible to maintain eye contact with the eyepiece. Unfortunately we did not have the proper connector to attach the pediatric scope to the video monitor. After discussing the case with the surgeon and the patient, the case was done under local anesthesia and sedation using mask ventilation of 0.6% of Sevoflurane and intermittent boluses of Ketamine. The patient maintained hemodynamically stable through out the procedure.

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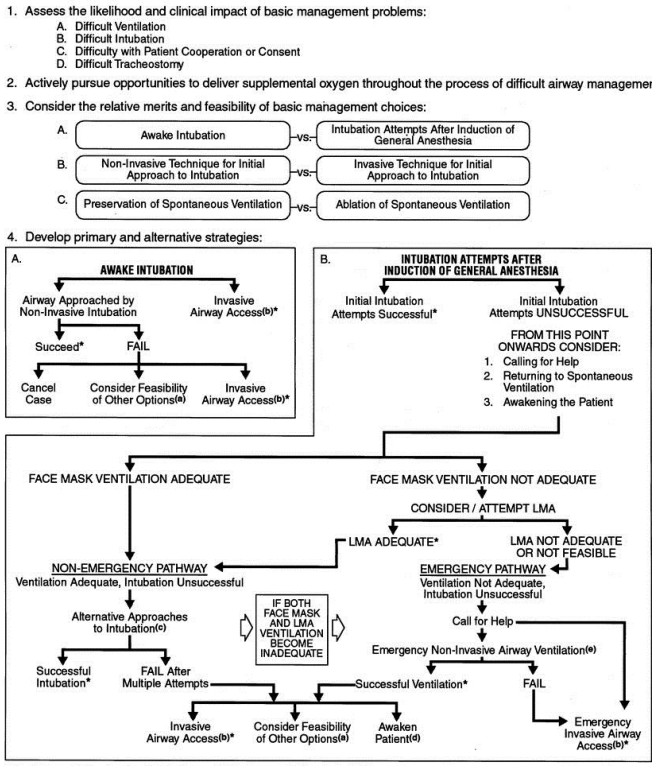

Figure 1: ASA Guidelines.

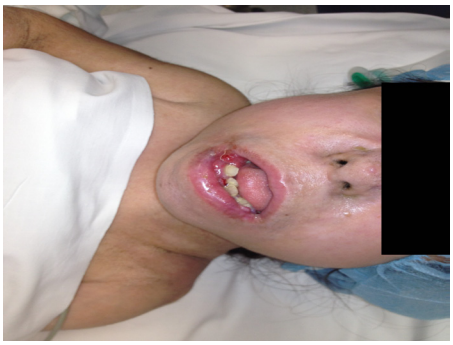

Figure 2: Examination of the airway.

Discussion

Airway management in patients with ISH can be quiet challenging for the anesthetist because of the clinical features of the disease. Predictors for difficult intubation in those patients includes: gingival hyperplasia, limited mouth opening, restricted neck movement, protrusion of the mandible all caused by immobility of the joints by progressive contractures. In addition, the presence of nodules along the airway can further complicate intubation by alteration of the normal anatomy. For ISH patients, direct visualization is mandatory when there is airway pathology. Planning and timing for any procedure that involves these patients is crucial and following the ASA guidelines in difficult intubation can be helpful [3] (Figure 2). In general, airway management can be approached by one of the following:

Laryngeal mask airway

In spite of limited mouth opening, trial of LMA had failed in this patient, most likely because of the airway nodules that prevented proper positioning of the LMA.

Oral intubation

1) Direct visualization with laryngoscope: is possible at early stages of the disease but once the disease advance and involves TMJ, it is almost difficult to visualize the vocal cords.

2) Glidescope/C-mac: is another option but limited mouth opening (<3 cm) is a relative contraindication, but it is worth trying.

3) Fibro-optic intubation: in our case, several attempts were done, but all failed. Having the pediatric fiber-optic attached to a monitor would have made a difference. Fibro-optic is operator dependable and the presence of an expert anesthetist is essential for the success.

Nasal intubation

Could be done through fibro-optic scope. In our case, the patient had very narrow nasal nares and the presence of nodules made it impossible. One case report by Norman and colleagues about 2 pediatric patients with ISH were successfully intubated using nasal intubation with fibro-optic because of limited mouth opening [4].

Retrograde intubation

Rarely done, but may be a good option in this case, however, it requires a co-operative patient and special preparations.

Tracheostomy

Least likely to be done, but it is the last resort for elective cases.

Recommendations

Because of the nature of the disease process, which tend to progress with age, a thorough airway examinations should be done for those patients to assess the difficulty of the airway. For most of these patients, mask ventilation is not a problem and it can be used for elective short procedures, where LMA cannot be used. Whereas in major surgical procedures that require intubation, then fiber-optic would be the best available option. In case of emergency where intubation fails, surgical airway is a must for those patients.

References

1. Al-Mayouf SM, AlMehaidib A, Bahabri S, Shabib S, Sakati N, et al. (2005) Infantile systemic hyalinosis: a fatal disorder commonly diagnosed among Arabs. Clin Exp Rheumatol 23: 717-720.

2. Shieh JTC, Hoyme HE, Arbour LT (2008) Hyalinosis, Inherited Systemic. In: Pagon RA, Bird TD, Dolan CR (eds). GeneReviews™. Seattle (WA) (1993): University of Washington, Seattle, USA.

3. Anesthesiology (2003) © 2003 American Society of Anesthesiologists, Inc. Lippincott Williams & Wilkins, Inc. Practice Guidelines for Management of the Difficult Airway. An Updated Report by the American Society of Anesthesiologists Task Force on Management of the Difficult Airway 98:1269-1277.

4. Norman B, Soni N, Madden N (1996) Anaesthesia and juvenile hyaline fibromatosis. Br J Anesth 76: 163-166.