A CASE OF ACHALASIA ON A DOWN SYNDROME PATIENT: BALANCING BETWEEN A PREDICTABLE DIFFICULT AIRWAY AND THE RISK OF ASPIRATION

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BACKGROUND AND GOAL OF STUDY

Achalasia is a neuromuscular disorder of the esophagus characterized by inadequate relaxation of the lower esophageal sphincter (LES) and loss of esophageal peristalsis, leading to an increased risk of massive regurgitation and aspiration of esophagogastric content. Its prevalence in Down Syndrome patients is superior to that of the general population. Besides, Down syndrome may occur with other structural anomalies associated with a possible difficult airway. Our aim is to shed a light on the anesthetic management of these patients.

CASE REPORT

PREANAESTHETIC ASSESSMENT

A 47-year-old male, **ASA physical status III**, was posted for Heller's myotomy due to achalasia. From the preanaesthetic assessment, we highlight:

Medical History

- Down Syndrome
- Achalasia with active regurgitation of esophagogastric content and loss of 40 kg in the last 2 years
- Corneal transplant

Airway examination

- Mallampati II
- Macroglossia
- Lack of teeth
- No history of prior difficult airway or stigmas of difficult ventilation

Fluoroscopy with barium swallow showed a typical bird beak sign with associated esophageal dilation (**figure 1**). The remaining preanaesthetic exams (routine blood investigations and EKG) were within the normal limits.

After taking informed consent, the patient was kept on a fasting diet on the night prior to the surgery.

INTRAOPERATIVE PERIOD

We used a **general anesthesia** as our strategy:

- Firstly, we inserted a naso-esophagic tube with the patient awake, for aspiration of the esophagagic content (approx. 100ml).
- A rapid sequence induction and intubation (RSII) with direct laryngoscopy and stylet was done, in a semi-sitting position (**figure 2**) with cricoid pressure. Endotracheal intubation was successful at first attempt.
- Intraoperative monitoring included ASA standards and BIS.

The patient remained hemodynamically stable throughout the surgery, that lasted for 3h35min. He was extubated successfully and admitted to the Post-Anesthesia Care Unit.

POSTANAESTHETIC CARE

In the **PACU**:

- The patient remained hemodynamically stable, eupneic, awake and pain-free.
- A multimodal analgesic approach was used for pain control.

The remaining postoperative period was uneventful and the patient was discharged 2 days after the surgery.

Figures: 1 – Bird-beak sign typical of esophageal achalasia; **2** – semi-sitting position for RSII

DISCUSSION

Achalasia is a chronic condition without cure. Current treatment options are aimed at reducing the hypertonicity of the LES. Heller's myotomy remains the best surgical treatment.

Patients with achalasia are at high risk of perioperative aspiration as their dilated esophagus may retain food for many days. Fasting period is meaningless and we must use full-stomach precautions.

As our patient had Down syndrome, the predictability of a difficult airway was an additive problem. The presence of several criteria for difficult airway should prompt a fiberoptic intubation, following the current guidelines. However, we found some limitations in this case, particularly the need for a deeper sedation in a patient that didn't cooperate due to cognitive dysfunction and the important risk of massive regurgitation.

We opted to decompress and empty the esophagus prior to induction with a large-bore nasogastric tube and perform a RSII with a stylet and cricoid pressure in semi-sitting position. It was a successful approach that enabled an eventless perioperative period for this patient.

LEARNING POINTS

Anaesthetic management of patients with a predicted difficult airway for surgical treatment of achalasia is very challenging. Suitable pre-anaesthetic preparation for difficult airway management is the key.

Fiberoptic intubation may not be the best option for a difficult airway in a uncooperative patient with high risk of massive regurgitation.