AIRWAY MANAGEMENT: A PATIENT WITH MADELUNG DISEASE

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This case study describes a 51-year-old, ASA physical status II Hispanic man who underwent liposuction to the anterior neck and resection of adipose tissue of the posterior neck. The patient was diagnosed with multiple symmetric lipomatosis, better known by its eponym, Madelung disease. This disease is characterized by abnormal adipose tissue growth primarily in the neck and shoulder regions. As a result of this pathology, a complete airway assessment was challenging. The patient’s Mallampati classification was class III with visualization of the soft palate only.

Other elements of a full assessment such as thyromental distance, sternomental distance, and tracheal orientation were impossible to evaluate because of the extreme circumferential girth of the adipose tissue in the neck. In addition, flexion and extension of the neck was limited by a minimum of 20° because of the pathology. The patient was deemed as having a difficult airway, and an awake fiberoptic intubation was planned and used successfully. The subsequent surgical procedure progressed without incident, and the patient was transferred to a stepdown unit for monitoring of his airway postoperatively.

Key words: Fiberoptic, lipomatosis, Madelung disease.

Case summary

The patient was a 51-year-old, ASA physical status III Hispanic man who was admitted for liposuction to the anterior neck and resection of adipose tissue of the posterior neck. The patient had a medical history of type 2 diabetes mellitus, obesity, and benign prostatic hypertrophy. The patient was 69 in tall and weighed 104.5 kg. His body mass index was calculated as 34. The diagnosis of Madelung disease was made when the patient was 31 years old. His surgical history revealed multiple liposuctions to the neck without tissue resection. Incidentally, the patient stated that there were no complications with his prior anesthetics, which included general anesthesia.

The patient's airway examination, using the Mallampati classification system, revealed a class III airway. The airway classification system is based on the idea that a disproportionately large tongue base could prevent exposure of the larynx. The inability to directly measure the tongue base leaves only an indirect measurement by noting the relative visibility of pharyngeal structures. The classes range from I to IV, with potential difficulty in airway management rising with each increase in number. The patient had a mouth opening distance of approximately 3 finger-breadths. An evaluation of the thyromental distance, sternomental distance, and tracheal orientation was impossible to evaluate because of the extreme circumferential girth of the adipose tissue in the neck. In addition, flexion and extension of the neck was limited by a minimum of 20° because of the pathology. The patient was deemed as having a difficult airway, and an awake fiberoptic intubation was planned and used successfully. The subsequent surgical procedure progressed without incident, and the patient was transferred to a stepdown unit for monitoring of his airway postoperatively.

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Multiple symmetric lipomatosis, or Madelung disease, is described in Harrison’s Principles of Internal Medicine as a lipomatosis that affects men 4 to 15 times more frequently than it affects women. Furthermore, it is characterized as a symmetric, progressive growth of nonencapsulated subcutaneous adipose tissue primarily in the neck and shoulder regions. Accumulation of fat in the trunk and proximal limbs may occur, but the legs and distal arms are left uninvolved. Laryngeal, tracheal, or vena caval compression can occur but is rare.

Most patients have a significant alcohol abuse history. However, the mechanisms and predisposing factors remain largely unknown. What is known is that the tissue contains small adipocytes with increased lipoprotein lipase activity and reduced catecholamine-stimulated lipolysis. It also has been reported that mitochondrial DNA mutations—A-to-G and G-to-A transitions—have occurred in some sporadic cases.

Expansion of adipose tissue mass via fat cell proliferation was termed hyperplastic obesity by Bjornorp and Sjostrom to separate it from hypertrophic obesity, in which increased fat cell size is thought to be the major process by which adipose tissue mass increases. It is thought that expansion of adipose tissue stores by fat cell proliferation, not fat cell hypertrophy, may be a more metabolically benign way of increasing energy stores.

Not all is known about Madelung disease, but it is clear that real challenges are presented to anesthesia providers by this patient population. This case study describes 1 patient and the interventions used in providing a safe anesthetic.
intubation and attempting to allay any preexisting fears, anxieties, and concerns the patient verbalized. The elements of comfort and safety provided with this procedure were stressed to the patient. To assist with anxiolysis, 2 mg of midazolam was titrated intravenously for sedation, and 0.3 mg of glycopyrrolate was administered intravenously for its antisialagogue effect. For aspiration prophylaxis, the patient was given 30 mL of sodium citrate, 20 mg of famotidine, and 10 mg of metoclopramide. Topicalization of the airway was performed via nebulization using 4 mL of 4% lidocaine.

To supplement topicalization, a glossopharyngeal nerve block was performed to blunt the gag reflex. The gag reflex is stimulated by deep pressure receptors in the posterior third of the tongue, which are difficult to block with diffusion of local anesthetics through the mucosa. The patient was transferred from the preoperative holding area to the operating room, and standard monitors were applied. All required anesthetic drug modalities, airway equipment, and anesthesia providers skilled in airway management were immediately available.

The patient was positioned in a beach chair position for placement of the glossopharyngeal nerve block. The patient was instructed to protrude his tongue anteriorly, and a tongue blade was used to displace the tongue medially. This approach assisted in the formation of an anatomical gutter along the floor of the mouth between the tongue and the teeth. The gutter ends in a cul-de-sac formed by the base of the palatoglossal arch. A 25-gauge spinal needle was inserted at the base of the cul-de-sac and advanced 0.5 cm. After a negative aspiration test, 2 mL of 1% lidocaine was injected. This technique was repeated on the contralateral side. Because the excessive adipose tissue prevented the identification of pertinent anatomical structures, a superior laryngeal nerve block was contraindicated. Likewise, blocking sensory innervation of the trachea and vocal cords by the recurrent laryngeal nerves, using a transtracheal technique, also was impractical.

The patient maintained spontaneous ventilation with an anesthesia face mask with a fraction of inspired oxygen of 100% for 5 minutes to implement denitrogenation. An additional 3 mg of midazolam and 100 µg of fentanyl was titrated intravenously to enhance amnesia and sedation respectively. During a 5-minute period of preoxygenation, the patient was relaxed, cooperative, and responsive. A 10-cm intubating oral airway was placed without evidence of a gag. A fiberoptic scope was advanced to the level above the vocal cords, and 5 mL of 2% lidocaine was deposited via the suction port of the fiberoptic scope using the “spray as you go” technique. The patient responded with mild coughing. The scope was passed...
through the vocal cords, and an 8.0 endotracheal tube was passed to 24 cm at the lip. The endotracheal tube cuff was inflated with 7 mL of air. Breath sounds were auscultated bilaterally, and greater than 4 positive end-tidal carbon dioxide reflections were noted on the monitor. After confirmation of endotracheal tube placement and securing of the tube, the patient was given 150 µg of fentanyl, 150 mg of propofol, and 100 mg of rocuronium. The patient then was placed on mechanical ventilation and was maintained under general endotracheal anesthesia using desflurane as the volatile inhaled agent.

The surgery progressed without incident. The patient initially was placed in a supine position to complete liposuction to the anterior and lateral aspects of the neck. After completion of liposuction to the anterior neck, the patient was turned prone in a careful and controlled manner for resection of the abundant adipose tissue on the posterior neck. The weight of the resected tissue mass was nearly 3 lb. The surgical procedure concluded with the approximation of skin at the posterior neck.

The patient met requirements for extubation at the end of the case and was extubated without incident. The patient recovered in the postanesthesia care unit, and, after an uncomplicated recovery, he was transferred to a stepdown unit for further critical care monitoring of his airway and hemodynamic status. The surgical insult was circumferential to the neck and posed great concern for postoperative airway compromise. After 24 hours, the patient was transferred to a surgical ward where his recovery was uneventful.

**Discussion**

Patients with Madelung disease have important airway considerations. As with any recognized difficult airway, preparation of the patient for an awake intubation is paramount. This begins with the psychological preparation of the provider and the patient. The anesthesia practitioner should be skilled and confident in the management of an awake intubation. Preparedness, which promotes self-assurance, includes a thorough review of previous medical records, health history, and a detailed airway assessment. The ability to communicate is vital because this allows the anesthesia provider to minimize the patient’s emotional distress while effectively sharing the plan with the surgical team.

The second area of significance is the safe use of premedication and intravenous sedation. The key is safely titrating enough medication for anxiolysis without creating an uncooperative patient who lacks the ability to guard his or her own airway. Such a scenario is counter-intuitive and unsafe when considering an awake intubation. Other important premedication regimens worthy of consideration include the use of antisialagogues and medications for aspiration prophylaxis.

The third area of concern is strategic preparation, which includes transport, staff, monitors, oxygen, and airway equipment. There is no time in crisis to be looking for needed equipment. All equipment should be immediately available and functional. Having the correct equipment and providing proper preparation is vital for a successful anesthetic within this patient population. All measures should be taken to ensure that a controlled system of airway management does not turn into a catastrophic event.

Finally, optimization of the patient’s airway with effective topicalization and the proficient use of airway nerve blocks will promote a smooth awake intubation. Not all blocks are feasible as demonstrated with this case study. However, a combination of techniques can be used to facilitate a successful intubation.

Although these areas of concern, which have been previously developed, are not particular to Madelung disease, they serve as a foundation when planning and implementing an awake intubation for any patient population. The goal is to provide the patient with the safest plan of care, while maintaining command and control of the situation.
REFERENCES

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