Congenital Subglottic Stenosis Complicating Tracheal Intubation in an Adult With Dwarfism

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Case Report

INTRODUCTION

Subglottic stenosis is a congenital or acquired narrowing of the subglottic airway, which can present as an unrecognized difficult airway (1). It is commonly associated with multiple intubations, prolonged intubation time, and infection. It may also be caused by a congenital condition occurring in the setting of dwarfism (2). A thorough history and physical exam with special attention to the airway may help us recognize this problem. However, in patients without a prior history of intubation, it may present as the inability to advance an endotracheal tube during intubation.

CASE REPORT

A 47-year-old male with past medical history (PMH) of dwarfism, arthritis, and degenerative spinal disease presented for revision total hip arthroplasty under general anesthesia (GA). Past surgical history included six joint surgeries, GA with laryngeal mask airway, and reported difficulty with neuraxial anesthesia. There were no records of previous intubations. Physical exam revealed short stature and limited neck extension from cervical spine disease. Airway exam demonstrated a Mallampati score of II, thyromental distance < 6 cm, small mouth opening, and a short neck.

GA was proposed with video laryngoscopy due to an assumed difficult airway secondary to inability to hyperextend atlanto-occipital joint and small mouth opening. A large epiglottis and large arytenoids with abducted vocal cords were visualized deeper than anticipated. A 6.5 mm endotracheal tube (ETT) was unable to be passed through the vocal cords despite multiple attempts. The surgery was postponed for ENT evaluation and imaging, which demonstrated a diffusely small caliber airway resulting in mild narrowing of the subglottis, an anterior shelf, and angulation of the anterior arch of the cricoid cartilage.

A second surgery was attempted, wherein a cuffed 5.5 ETT was inserted through the vocal cords, requiring some rotation to pass through the anterior subglottic shelf. The surgery was performed uneventfully and patient was extubated then transferred to PACU where he recovered without complication.

DISCUSSION

Subglottic stenosis (SGS) results from acquired or congenital narrowing of the subglottic airway (3). Acquired SGS is more common than congenital SGS, accounting for 95% of all cases. It is most commonly a result of endotracheal intubation and the coupled inflammatory response (4).
Congenital SGS is believed to be a malformation of the cricoid cartilage as a result of failure of the laryngeal lumen to recanalize properly during embryogenesis (5). Congenital SGS is associated with other congenital abnormalities and is divided into membranous and cartilaginous types. Cartilaginous SGS is usually caused by a deformed or hypertrophied cricoid cartilage. This cartilage has the ability to form an anterior subglottic shelf, which extends posteriorly and results in a small posterior opening (5).

Based on ENT evaluation, imaging, and the presence of the subglottic shelf, we believe we encountered cartilaginous type congenital SGS in our patient. Congenital SGS may be associated with several diseases, one of which is achondroplastic dwarfism (2,6). Atypical bone growth in achondroplastic dwarfism can be associated with possible anesthesia related complications (2).

Although difficult laryngoscopy was suspected in this case secondary to limited neck extension, small mouth opening, and short neck, difficulty intubating was not expected. Because it is known that early ossification occurs in achondroplastic dwarfs (7), we hypothesize that ossification of the cartilaginous portion of the larynx further complicated passage of the ETT worsening the patient’s subglottic stenosis. This provides further evidence of the relationship of subglottic stenosis with achondroplastic dwarfism.

CONCLUSION

We encountered unrecognized subglottic stenosis that led to difficult tracheal intubation in a patient with achondroplastic dwarfism. In patients with dwarfism, the anesthesiologist must be aware of the possibility of congenital subglottic stenosis. Preoperative ENT evaluation and imaging of the airway prior to attempting endotracheal intubation or general anesthesia may diagnose problems that could arise during intubation and help mitigate intra- and postoperative complications.

SOURCES


