Anesthetic Management of a Juvenile Hyaline Fibromatosis Patient With Trismus and Cervical Movement Limitation

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Juvenile hyaline fibromatosis (JHF) is a rare autosomal recessive disease characterized by the presence of tissue nodules, joint contractures, and gingival hyperplasia. With a 1-year-9-month-old female patient scheduled for a gingivectomy and excision of a lower lip mass under general anesthesia, it was anticipated that airway management would be difficult because of trismus and limited cervical movement. Intubation with video-laryngoscopic assistance could not be achieved because gingival hyperplasia and trismus prevented blade insertion and manipulation. Therefore, 2 endotracheal tubes were used: 1 used as a nasopharyngeal airway for assisted ventilation, and 1 used for intubation along with a flexible fiberoptic scope. This case demonstrated a useful method for managing ventilation and intubation in patients with JHF, particularly when the use of oral airway devices is difficult.

Key Words: Juvenile hyaline fibromatosis; Trismus; Difficult intubation; Fiberoptic scope; Nasotracheal intubation.

Juvenile hyaline fibromatosis (JHF) is a rare genetic disease involving mutations of the ANTXR2 gene causing the accumulation of hyaline substance in various tissues and is characterized by multiple subcutaneous nodules or masses, gingival hyperplasia, and joint contracture. Severity of signs and symptoms occur on a spectrum with more severe cases typically occurring at or shortly after birth.[1–3]

The patient was a 1-year-9-month-old female (height 76.5 cm; weight 7.2 kg) who was previously hospitalized twice with pneumonia. She was scheduled for a gingivectomy and excision of a lower lip mass under general anesthesia. Difficulties maintaining the airway during anesthetic management were anticipated due to trismus (15 mm between the upper and lower primary central incisors), limited cervical movement, and gingival hyperplasia. The patient's airway was easily maintained after slow mask induction with sevoflurane and oxygen. Adequate mask ventilation was confirmed, and the following drugs were intravenously administered: 0.08 mg atropine, 15 μg fentanyl, and 5 mg rocuronium bromide. Thereafter, continuous infusions of propofol (14 mg/kg/h) and remifentanil (0.2 μg/kg/min) commenced. Endotracheal intubation was attempted with a video laryngoscope (AIRWAY SCOPE, PENTAX) fitted with a thin blade (M-ITL-PL, PENTAX) designed for pediatric use. However, insertion and maneuvering of the blade was quite limited due to trismus and gingival hyperplasia and caused difficulty observing the glottis. Hence, the video laryngoscope was changed out for a flexible fiberoptic scope along with the placement of 2 standard-shaped oral/nasal endotracheal tubes (ETTs): a cuffless 4.0 mm ETT (Covidien) inserted into the left nasal cavity for assisted ventilation, and a Microcuff 4.0 mm ETT (Halyard Healthcare) inserted into the right nasal cavity for fiberoptic intubation. In addition, tape was used to cover the mouth and prevent air leakage, resulting in improved ventilation. Due to difficulty visualizing the glottis, fiberoptic intubation required 4 attempts for successful completion. Following surgery, a 4.0-mm nasopharyngeal airway was prophylactically inserted through the left nostril to help maintain airway patency. Extubation was performed

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carefully once the patient was awake with the use of a tube exchanger inserted through the ETT and into the trachea. After confirming stability of the patient's respiratory status postextubation, the tube exchanger and nasopharyngeal airway were removed.

JHF is a rare autosomal recessive disease first reported by Murray.[1] Maintaining the airway during anesthesia can be difficult due to masses within the oral cavity and trachea, as well as temporomandibular and cervical joint contracture.[4–6] Although the Airway Management Guidelines by the Japanese Society of Anesthesiologists[7] recommend conscious intubation if difficulty in maintaining the airway is expected, it is difficult to follow this recommendation when treating an infant or young child. Here a video laryngoscope fitted with a thin blade specifically for pediatric use was selected as the first-line choice. However, due to difficulty in observing the glottis, it was changed to nasal fiberoptic intubation. It can be difficult to achieve sufficient respiratory volumes when performing both manual ventilation and fiberoptic intubation through a single ETT in a pediatric patient due to the small size of the tracheal tube so a second ETT was placed. Considering the possible difficulty with using oral airway devices for a patient with JHF with gingival hyperplasia and trismus, this method of nasal fiberoptic intubation with nasopharyngeal assisted ventilation may be the ideal first-line method.

Additionally, the deposition of hyaline substance during the progression of JHF can cause stenosis of the nasal cavities and the trachea.[5] It is advisable to consider utilizing a computed tomography scan when preoperatively assessing the airway (ie, the laryngopharynx and nasal cavities) for the presence of stenosis. Additionally, it is important to note the possibility for emergent reintubation when extubating a patient with JHF. In the present case, in addition to difficulties with intubation, the potential for emergent reintubation due to hemorrhage after gingivectomy could not be ruled out, so a tube exchanger was utilized.

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