Difficult Airway Management in Osmed Syndrome

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Dear Editor;

Otospondylomegapiphyseal dysplasia (OSMED) is an autosomal recessive inherited disorder characterized by sensorineural hearing loss and short legs associated with enlarged epiphyses (1). It is a rare disease, and many mutations and polymorphisms, such as micrognathia, glossoptosis, nonsyndromic cleft palate, Kawasaki disease, coronary artery lesions and decreased hearing, are reported in the COL11A2 gene (2). Typical facial features include mid-facial hypoplasia with a small nose and a flattened bridge of the nose. We wanted to share difficult airway management for a patient who was scheduled for nasal reconstruction with rib grafting due to flattened bridge of the nose (saddle nose) and operated.

A 21-year-old patient had sensorineural hearing loss, micrognathia, protruding upper incisors, proximal cortical thickening and anterior curvature of both tibia (Figure 1). Anaesthesia induction with fentanyl and propofol after midazolam

Figure 1. Presence of micrognathia, flattened nose and protrusive eyes in accordance with OSMED syndrome

premedication and routine monitoring and muscle relaxation were provided. The patient, who had not been intubated in three attempts in spite of cricoid compression with a stylet and positioning by elevation of the head (Mallampati score III, Cormack-Lehane score IV) was hardly intubated with a video laryngoscope. At the end of the operation, lasting for approximately 80 minutes, the patient was extubated with sugammadex and referred to the service after a while.

In these patients, palate and mid-facial congenital anomalies show a high possibility of difficult intubation (3). In this condition, the preoperative evaluation and difficult airway preparation must be complete. Moreover, airway instruments, including a laryngeal mask, flexible fiberoptic bronchoscope and video laryngoscope, must be kept available.

In our case, the characteristic findings and typical facial appearance, such as disproportionate short stature, short lower extremities and enlarged epiphyses, vertebral body abnormalities and sensorineural hearing loss, were remarkable. The medical history of the patient was questioned in detail, and no pathology was detected in the cardiological examination.

Awakening and recovery can also be difficult in patients who had difficult intubation (4). Short-acting rocuronium was preferred for its rapid myorelaxant action, and the specific antagonist sugammadex, which is able to reverse the myorelaxant activity in a prompt fashion. Thus, delayed extubation and haemodynamic fluctuation were prevented. In OSMED syndrome, preoperative evaluation and preparations in the operating room are important with regard to mortality and morbidity because of a difficult airway. We thought that

sharing the typical findings in our case and the benefits of alternative airway instruments in the management of difficult airways would be helpful to your readers.

Informed Consent: We informed the patient, that we may publish the medical information and photos from the operation, without harming the patient's anonymity.

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