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LETTER TO THE EDITOR

Management of general anesthesia for a patient with Maroteaux type acromesomelic dysplasia complicated with obstructive sleep apnea syndrome and hereditary myopathy

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To the Editor:

Oropharynx abnormality may lead to difficult intubation in patients with osteochondrodysplasia [1]. For the very rare disease, acromesomelic dysplasia Maroteaux-type (AMDM), which is one such abnormality, however, there is no report whether special consideration is needed in airway management. AMDM is seldom combined with other significant disease [2]. We report the first case of a patient with AMDM complicated by severe obstructive sleep apnea syndrome (OSAS) and hereditary myopathy undergoing smooth management of general anesthesia for elective L4–S1 laminectomy and discectomy.

AMDM, which was first described by Maroteaux et al. in 1971 [3], is a very rare autosomal recessive disorder of progressive osteochondrodysplasia with a prevalence of about 1 in 2,000,000 [2]. It is characterized by severe dwarfism and acromesomelia, which presents with disproportionately shortened middle and distal segments of the four limbs.

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The 33-year-old female patient suffered from AMDM, which was diagnosed 13 years ago based on gene mapping (Affymetrix Genechip Mapping 100K set), in which the linkage data showed abnormality in chromosome 9 [4], and on typical clinical features, such as short stature with disproportionately shortened distal part of limbs (Fig. 1). She also had severe OSAS with apnea-hypopnea index up to 48, and had received treatment of bi-level positive airway pressure for 3 years [5]. Muscle biopsy performed 2 years ago in response to progressive muscle weakness had suggested hereditary muscle atrophy, which was not considered AMDM related.

Preoperative evaluation revealed no significant head deformity except prognathia. Neck range of motion was normal. Mallampati classification was grade 2. Muscle power of the four limbs was four, worse in the distal parts. Preoperative laboratory data, chest X-ray, and pulmonary function test were unremarkable.

Under train-of-four (TOF) monitoring over the adductor pollicis, induction agents of fentanyl 100 μg, 2 % lidocaine 40 mg, propofol 100 mg, and cisatracurium 7 mg were given intravenously after sufficient preoxygenation. When the patient was apneic, an oropharyngeal airway was necessary for mask ventilation. When the second twitch of TOF disappeared, endotracheal intubation using direct laryngoscope was performed uneventfully under the view of Cormack and Lehane classification grade one. After a 2-h surgical procedure, tracheal extubation was performed under fully awake conditions and recovery of TOF in the operating room. The patient was then discharged smoothly on the fourth postoperative day without any sequelae of general anesthesia.

As this is a very rare case report, we do not know whether patients with AMDM may have a difficult airway. In such a situation, succinylcholine may be the first choice

Fig. 1 The 33-year-old female patient with acromesomelic dysplasia Maroteaux-type (AMDM) was of typical short stature, 126 cm in height, weight 36 kg, and had disproportionately shortened forearms, wrists, fingers, legs, and toes (acromesomelia). (Photograph reprinted with permission obtained from the patient)

of muscle relaxants for induction of general anesthesia. Nevertheless, it was not the case for this patient because of underlying hereditary myopathy. In conclusion, we report a very rare case of AMDM complicated with hereditary myopathy and OSAS undergoing general anesthesia with cisatracurium smoothly. In this case, AMDM itself seems not to complicate airway management during intubation or extubation.

References

1. Hattab FN, Al-Khateeb T, Mansour M. Oral manifestations of severe short-limb dwarfism resembling Grebe chondrodysplasia: report of a case. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1996;81(5):550–5.
2. Olney RC. C-type natriuretic peptide in growth: a new paradigm. Growth Horm IGF Res. 2006;16(suppl A):S6–14.
3. Maroteaux P, Martinelli B, Campailla E. Le nanisme acromésomélique. Presse Med. 1971;42:1839–42.
4. Sarina GK, Alexander P, Stefan M, Bernhard Z, Ralph TT, Harmien MZ, Shih LY, Arie H, Matthew LW. Acromesomelic dysplasia Maroteaux type maps to human chromosome 9. Am J Hum Genet. 1998;63:155–62.
5. Gross JB, Bachenberg KL, Benumof JL, Caplan RA, Connis RT, Coté CJ, Nickinovich DG, Prachand V, Ward DS, Weaver EM, Ydens L, Yu S. American Society of Anesthesiologists Task Force on Perioperative Management. Practice guidelines for the perioperative management of patients with obstructive sleep apnea: a report by the American Society of Anesthesiologists Task Force on Perioperative Management of patients with obstructive sleep apnea. Anesthesiology. 2006;104(5):1081–93.