Case Report

Airway management of a child with frontometaphyseal dysplasia (Gorlin Cohen syndrome)

Anuradha Ganigara, Madhavi Nishtala, Yabagodu Rama Vakoda Chandrika, Kunigal Ravishankar Chandrakala

Department of Anesthesiology, Indira Gandhi Institute of Child Health, Bangalore, Karnataka, India

Abstract

Frontometaphyseal dysplasia (FMD), also called Gorlin-Cohen syndrome, is a rare hereditary X-linked dominant craniotubular bone disorder. The presentation describes the airway management of a 2-year-old child suffering from FMD with significant retrognathia, posted for major long bone corrective osteotomy. Induction with a combination of dexmedetomidine and ketamine preceded a successful endotracheal intubation under spontaneous ventilation.

Key words: Dexmedetomidine, difficult airway, frontometaphyseal dysplasia, ketamine

Introduction

Frontometaphyseal dysplasia (FMD), also called Gorlin Cohen syndrome, is a hereditary X-linked dominant syndrome described in 1969 with less than 30 cases described in the literature.[1,2] This case report of a child with FMD is presented owing to the rarity of the syndrome and the anticipated difficult airway, which was successfully managed by using a combination of dexmedetomidine and ketamine while preserving spontaneous ventilation.

Case Report

A 2-year-old female child, a known case of FMD, presented for open reduction of the left hip with osteotomy of femur. Physical examination revealed a slender undernourished girl of 8 kg with prominent supraorbital ridges, ocular hypertelorism, low set ears and a wide bridge nose with prominent eyes. Airway examination revealed a Mallampatti score of III with significant retrognathia, high arched palate with malocclusion of teeth. In addition, she had dorsolumbar scoliosis, pectus carinatum, bowing of long bones with distal phalangeal hypoplasia and multiple joint dislocations [Figure 1].

Pre-operative blood investigations, echocardiography and chest X-ray were within physiological limits. Oral midazolam 4 mg was selected for premedication. On arrival to the operation theatre, monitors were connected and child pre-oxygenated for 5 minutes.

Injection dexmedetomidine 1 μg/kg was administered for 10 min and then a continuous infusion at 1 μg/kg/h was set for the duration of the remaining procedure. Ketamine was administered in increments of 5 mg up to 12 mg until there

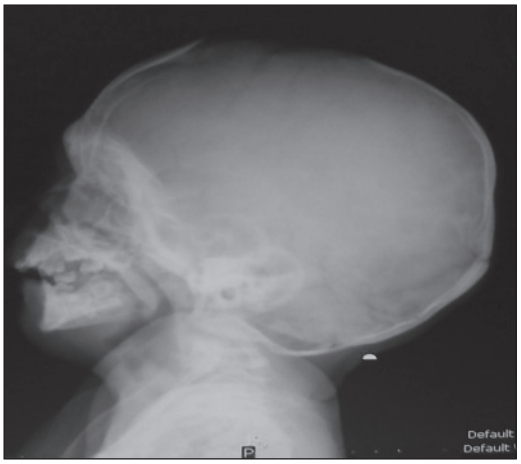

Figure 1: X-ray lateral view

Address for correspondence: Dr. Anuradha Ganigara, Department of Anesthesiology, Indira Gandhi Institute of Child Health, South Hospital Complex, Dharmaram College Post, Bangalore - 560 029, Karnataka, India.
E-mail: anuradhasudhir@gmail.com

Access this article online	
Quick Response Code:	Website: www.joacp.org
	DOI: 10.4103/0970-9185.130100

was no response to jaw thrust while ensuring spontaneous respiration. Just before direct laryngoscopy intravenous lignocaine 12 mg was administered. Rigid laryngoscopy with Miller 1 straight blade offered a Grade IV Cormack and Lehane view. An additional dose of ketamine 5 mg was administered. After optimal external laryngeal manipulation, the visible glottic chink was sprayed with topical lignocaine and tracheal intubation was successfully performed using an uncuffed 4 sized endotracheal tube. Anesthesia was continued with N_2O in 40% O_2 along with a continuous Dexmedetomidine and atracurium infusion. Adequate padding was provided at pressure points and extreme caution was exercised during positioning. At the end of the surgery tracheal extubation was successful.

Discussion

FMD belongs to the otopalatodigital spectrum syndromes that includes four phenotypically related conditions, otopalatodigital syndrome Types 1 and 2, FMD and Melnick-Needles syndrome.[1,2]

The most common manifestations include supraorbital hyperostosis, hypertelorism, down-slanting palpebral fissures, broad nasal bridge and micrognathia with anomalies of teeth and generalized skeletal dysplasia. Congenital heart disease, subglottic tracheal narrowing and genitourinary anomalies, muscular hypotonia.[3,4]

Micrognathia, microstomia and malocclusion of teeth may make direct laryngoscopy impossible; therefore, a well-planned airway strategy is mandatory.

Ketamine combined with dexmedetomidine was selected for induction of anesthesia. Ketamine was preferred in our case of anticipated difficult airway due to it's inherent sympathomimetic actions devoid of respiratory depression alongwith provision of excellent analgesia and amnesia.[5-7] Dexmedetomidine a specific and selective $\acute{\alpha}_2$-adrenoceptor agonist known for its sedative, anxiolytic, analgesic properties was used to complement ketamine. At the same time dexmedetomidine offsets the sympathomimetic effects of ketamine, also decreases the sialorrhea and emergence delirium associated with ketamine.[8,9] This unique pharmacological combination in the present case preserved the respiratory drive, allowed maintenance of a patent airway and provided sufficient sedation, analgesia and anesthesia to allow successful airway control. In addition, topical lignocaine was used as per recommendation of Aroni *et al.* [5] which states that ketamine does not depress coughing or swallowing reflexes.

Available literature describes the use of combination of both these drugs in children during procedural anesthesia and not as a complete anesthesia protocol in a challenging case.[10] The present experience of using this combination successfully paves the way to emerging new solutions for management of a difficult pediatric airway.

Hence safety profile, rapid onset of action with adequate sedation and analgesia provided by the ketamine and dexmedetomidine make them a distinctive drug combination in the pediatric difficult airway situation in a child with FMD.

Acknowledgment

We thank pediatric orthopedic surgeons of Indira Gandhi Institute of Child Health, Bangalore: Dr. Rudraprasad and Dr. Kiran Rajappa for their invaluable support and cooperation.

References

1. Gorlin RJ, Cohen MM Jr. Frontometaphyseal dysplasia. A new syndrome. Am J Dis Child 1969;118:487-94.
2. Jones KL. Smith's Recognizable Patterns of Human Malformation. 6th ed. Philadelphia, Pennyslavania: Elsevier Saunders; 2006. p. 450-2.
3. Robertson SP. Otopalatodigital syndrome spectrum disorders: Otopalatodigital syndrome types 1 and 2, frontometaphyseal dysplasia and Melnick-Needles syndrome. Eur J Hum Genet 2007;15:3-9.
4. Mehta Y, Schou H. The anaesthetic management of an infant with frontometaphyseal dysplasia (Gorlin-Cohen syndrome). Acta Anaesthesiol Scand 1988;32:505-7.
5. Aroni F, Iacovidou N, Dontas I, Pourzitaki C, Xanthos T. Pharmacological aspects and potential new clinical applications of ketamine: Reevaluation of an old drug. J Clin Pharmacol 2009;49:957-64.
6. Roelofse JA. The evolution of ketamine applications in children. Paediatr Anaesth 2010;20:240-5.
7. Jamora C, Iravani M. Unique clinical situations in pediatric patients where ketamine may be the anesthetic agent of choice. Am J Ther 2010;17:511-5.
8. Tobias JD. Dexmedetomidine: Applications in pediatric critical care and pediatric anesthesiology. Pediatr Crit Care Med 2007;8:115-31.
9. Iravani M, Wald SH. Dexmedetomidine and ketamine for fiberoptic intubation in a child with severe mandibular hypoplasia. J Clin Anesth 2008;20:455-7.
10. Tobias JD. Dexmedetomidine and ketamine: An effective alternative for procedural sedation? Pediatr Crit Care Med 2012;13:423-7.

How to cite this article: Ganigara A, Nishtala M, Chandrika YV, Chandrakala KR. Airway management of a child with frontometaphyseal dysplasia (Gorlin Cohen syndrome). J Anaesthesiol Clin Pharmacol 2014;30:279-80.
Source of Support: Nil, **Conflict of Interest:** None declared.