Case Report

Airway management of a child with mucopolysaccharidosis undergoing cervical spine surgery: A case report

ABSTRACT

"Mucopolysaccharidosis" (MPS) is a rare, autosomal recessive lysosomal storage disease characterized by deficiencies in 11 different lysosomal enzymes involved in the metabolism of glycosaminoglycans (GAGs) leading to its accumulation, the condition which results in anatomic abnormalities and multi-organ dysfunction that increases the risk of anesthesia complications. The patterns of accumulation form the basis of MPS classification into seven types of progressive diseases. Most of the MPS types have facial and oral characteristics that increase the risk of airway management. We are reporting a case of MPS with a challenging airway, an 11-years-old boy diagnosed with MPS type VI with a prominent facial character planned for cervical spine fixation versus decompression, successfully managed with fiberoptic bronchoscopy (FOB) guided by video laryngoscopy (VL).

Key words: Airway management, Mucopolysaccharidosis, Spine surgery

Case Report

Eleven-years-old boy, diagnosed with MPS type VI, was manifested with hydrocephalus, cervical stenosis, facial puffiness, mitral regurgitation, recurrent pneumonia, and musculoskeletal deformities [Figure 1a-c] and admitted for cervical fixation versus decompression levels C1 and 2.

Beside anesthesia team, ENT and cardiothoracic staff were oriented to be ready for interfering at any time during the procedure, all airway management tools, emergency drugs, and measures were prepared. Airway topicalization with lidocaine 10% nebulized for 20 min before induction, 100% O2 face mask maintained, all vital signs were normal, midazolam 1 mg and fentanyl 20 mcg i.v. administered, followed by sevoflurane 8% inhalation induction, and mask ventilation was maintained while easy bagging. The first trial with GlideScope blade-2 shows a large epiglottis, hardly a lower portion of the larynx was seen, and intubation was unsuccessful. As bag mask ventilation was easy, rocuronium 10 mg given i.v., trachea was intubated with 5.5-mm tube using FOB guided by Glidescopy. GA was maintained by total intravenous anesthesia (TIVA) in the form of propofol 150 and remifentanil 0.05 mcg/kg/min. After the end of surgery, sugammadex 2mg/kg i.v. before extubation using an 11-Fr cook tube exchanger and transferred to PACU.

Discussion

In MPS disorder, the abnormal storage of GAGs in connective tissues and bones may compromise the airway.[1] A varying

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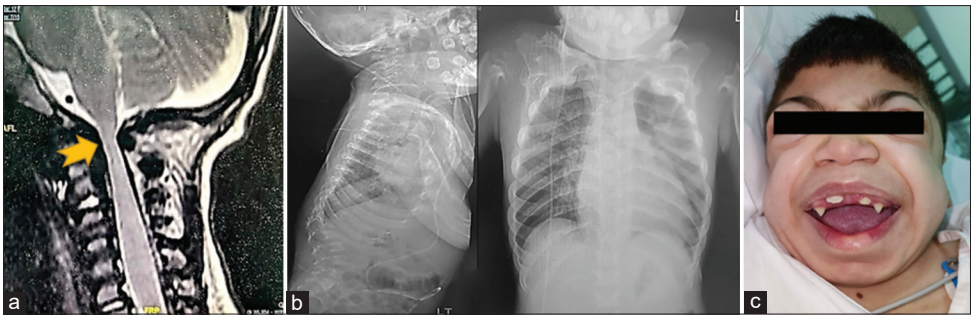

Figure 1: (a) Cervical stenosis, (b) CXR lateral and PA views, (c) Facial feature with M-IV and limited mouth opening

degree of airway obstruction was detected via endoscopy and imaging modalities described by Yi-Hao Lee, *et al*.[1] The data of 15 MPS cases were reviewed in detail. FOB was used to distinguish adeno-tonsillar hypertrophy, prolapsed soft palate, secondary laryngomalacia, vocal cord granulation, cricoid thickness, tracheal stenosis, shape of tracheal lumen, nodular deposition, tracheal kinking, tracheomalacia with rigid tracheal wall, and bronchial collapse. Up to 85% of these cases have reactive airway disease/asthma and sleep disorders.[2]

Brittney M. Clark, *et al*.[3] in 2018 had a comprehensive review of the literature with an emphasis on airway management for patients with MPS, they reviewed the literature for reports of perioperative course and airway-related anesthetic complications in patients with MPS, using a wide databases search strategy (1946–present), they identified nine case series, 27 individual case reports, and their airway management was summarized. In addition, case series of MPS patients who underwent anesthesia at Mayo Clinic between 2000 and 2015, which was reported in the Canadian Journal of Anesthesia, they described 18 MPS patients who underwent 49 procedures, 25% of them were intubated with VL. The overall incidence of difficult tracheal intubation ranges between 28% and 44% and intubations in MPS III patients was less difficult. VL was used as a rescue technique in patients who failed the initial planned approach.[3]

Megens *et al*.[4] reviewed the success rate of tracheal intubation using different tools: Direct laryngoscopy (DL) was difficult in 16 out of 55 cases, VL was successful in eight out of nine, and FOB intubations were performed without difficulty in only two out of ten cases. They reported an occurrence similar to our case, where a 15-year-old girl with MPS type VI, but she had difficult mask ventilation, and three failed attempts to place even a laryngeal mask airway, intubation was successful only with FOB aided by VL. The FOB was used by a second anesthetist to locate the glottic opening while VL provided only a view of the epiglottis.[4]

Conclusion

It is obvious that patients with MPS have a significant challenge for anesthetic airway management; however, the more widespread availability of VL made it the preferred technique for endotracheal intubation in such patients.[5] Anesthetic management should be in a well-equipped high specialized center.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images (by his father) and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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