10.5005/jp-journals-10029-1009

CASE REPORT

Anesthetic Management of a Case with Moebius Syndrome

Hande Arpaci, Merve N Kadioglu, Aysegul M Tuzuner-Oncul

ABSTRACT

Moebius syndrome is neurological disease of unknown etiology which is characterized by bilateral and unilateral facial and abducens nerve congenital paralysis. This syndrome usually presents with intubation difficulty due to the craniofacial anomaly and surgical interventions are required for treatment of inadequate chewing, swallowing, coughing reflex and hypotonicity, aspiration and respiratory problems. The aim of this case report is to present anesthesia technique during the extraction of 19 tooth in 2.5-year-old girl with severe hypotonia and possible difficult intubation due to craniofacial anomaly.

Keywords: Moebius syndrome, Sedoanalgesia, Tooth extraction.

How to cite this article: Arpaci H, Kadioglu MN, Tuzuner-Oncul AM. Anesthetic Management of a Case with Moebius Syndrome. Int J Exper Dent Sci, 2012;1(1):37-39.

Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

Moebius syndrome is an extremely rare congenital neurological disorder which is first described in 1888[1] and characterized by bilateral and unilateral facial and abducens nerve congenital paralysis.[2] The characteristics of patients having Moebius syndrome include: Possible accompanying mental retardation,[3] the involvement of V, IX, X, XI and XIIth cranial nerves which can lead to insufficiency in the chewing, swallowing, coughing reflex and respiratory problems due to aspiration.[4] Craniofacial anomalies, lip problems, musculoskeletal malformations, multiple ophthalmic anomalies,[5-7] involving seizure disorders, hypotonia and hypogonadothyropic hypogonadism[8] may occur with this syndrome.

The aim of this case report is to present the anesthesia technique during the extraction of the 19 tooth in 2.5-year-old girl with severe hypotonia, inadequate mouth opening and swallowing reflex, significant micrognathia, thyromental distance limitation and possible of difficult intubation because of the craniofacial anomaly.

CASE REPORT

A 2.5-year-old girl with Moebius syndrome referred to the Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, University of Ankara. General anesthesia was scheduled for the 9.5 kg weighted patient in order to prevent agitation and provide mouth opening for multiple tooth extraction surgery. Physical examination revealed; retrognathia, significant micrognathia, short philtrum, (Figs 1 and 2) deep palate, hypotonicity, low muscle tone, particularly in the upper body, inability to smile, strabismus (crossed eyes), bilateral epicanthus, inability of the eyes to abduct beyond midline, hirsutism in sacrum, presacral staining, limb abnormality, clubbed feet, missing fingers or toes. Bleeding-coagulation tests and hemogram-biochemistry parameters were found normal. The patient was not using any drugs for any treatment. It was reported that the patient was fed by an orogastric feeding tube till the operation day.

After 6 hours of fasting, the patient was taken into the operation room and monetarized (Mindray noninvasive vital

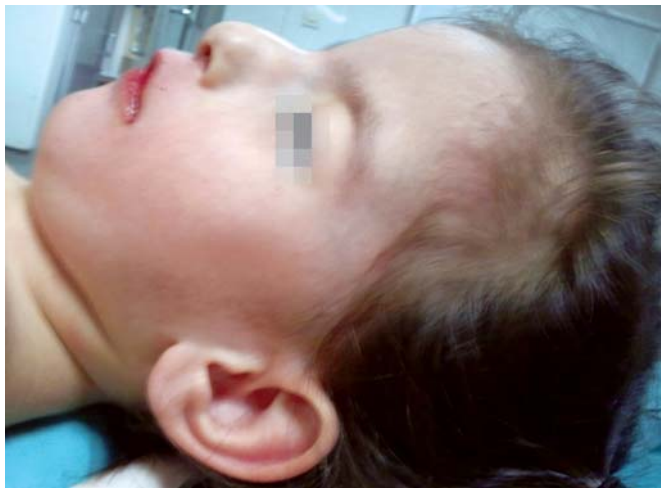

Fig. 1: Profile appearance of the patient

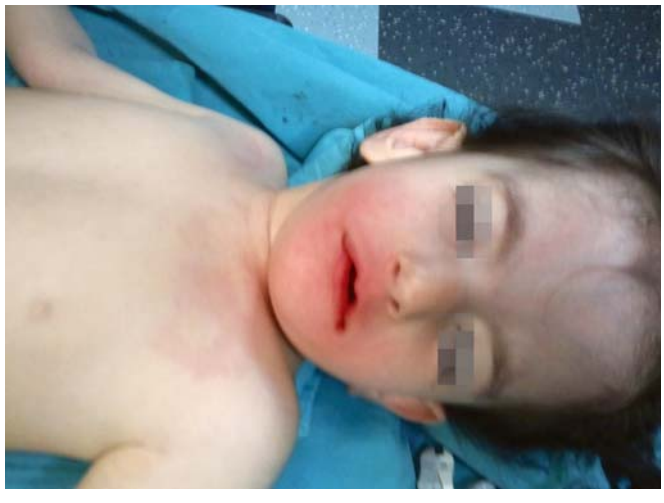

Fig. 2: Frontal appearance of the patient

sign monitoring system, MEC-2000) for the purpose of measuring noninvasive tension, peripheral oxygen saturation (SpO_2) and hearth rate with electrocardiography (KAH). After lidocaine HCL- prilocaine (Emla®) was applied on right hand dorsal side for topical local anesthesia, peripheral vascular canula was administrated.

As a result of inadequate mouth opening, micrognathia, retrognathia, thyromental, hyomental distance limitations in the patient, mallampati grade 4[9] as difficult intubation was predicted and nasal oxygen in a flow of 4 ml/min was administered. The hemodynamic parameters of the patient were stable, so 2 mg/kg ketamine (Ketalar®) for sedation with 5 mcg/kg atropine as an antisialagogue agent were applied. 120 mg paracetamol supozituar (Paranox Supozituvar®) for per-postoperative analgesia was administered following ketamine. The oxygen saturation was 97 to 100%, heart rate was 100 to 120 bpm, and mean blood pressure was 95 to 85 and 55 to 40 mm Hg during the operation period of 30 minutes. Additional ketamine with a dose of 0.5 mg/kg was applied at 10th and 20th minutes. Adequate mouth opening was maintained with ketamine and 19 teeth were extracted without application of local anesthesia. After suppository paracetamol administration, additional analgesia was not demanded. Three minutes after the operation was completed, the patient was waked up with a tactile stimulation. The patient was monitorized for 2 hours in recovery room, later with an adequate and regular respiration, the patient was discharged from the unit/hospital.

DISCUSSION

Moebius syndrome is a syndrome affecting multiple systems, having defects in extraocular eye movements due to congenital paralysis with single-sided or double-sided facial (VII) and abducens (VI) nerve and manifestations of craniofacial anomalies.[10] The etiology of Moebius syndrome is not fully understood;[11] an autosomal dominant, autosomal recessive and X-linked recessive inheritance have been reported but in cytogenetic examinations; on the 3rd and 13th chromosomes (3q21-q22 and 13q12.2-q13), two potential genetic loci were identified.[12-14]

The cases with Moebius syndrome frequently need anesthesia for imaging or correction of eyes anomalies (diplopia, ptosis operations, etc.), orthopedic problems (correction of limb abnormalities), plastic/reconstructive, otolaryngological, dental and general surgical intervention.[15]

Hypoplastic tongue, micrognathia, microstomia, cleft palate, bifid uvula, craniofacial abnormalities, such as ear abnormalities causing hearing problems, are observed quite frequently at cases with Moebius syndrome and these findings may lead to difficult intubation.[5] Ames et al reported that anesthesia was applied to 46 cases with Moebius syndrome, difficult intubation was the problem in seven of the patients and the intubation could not be done in one patient.

In this present case with the prediction of difficult intubation, the operation room was prepared for the algorithm for difficult airway, and the sedoanalgesia procedure was begun.

One of the most common findings associated with Moebius syndrome is hypotonia. Hypotonia can also affect postoperative respiratory function at both the level of the upper airway and the thoracic musculature and diaphragm. And especially in perioperative period, insufficient metabolization of anesthetic agents and muscle relaxants can lead to the development of respiratory failure.[4] Hypoventilation and respiratory failure were frequently observed following necrosis or secondary hypoplasia of pons and medulla in the children with Moebius syndrome.

Nineteen tooth extractions were planned in the patient who was diagnosed with Moebius syndrome after several procedures. She was followed up for 26 days in newborn intensive care unit because of the perinatal asphyxia. Having dissemine hypotonia, abnormal eye movements, feeding problems due to difficulty in swallowing the patient had applied to a hospital, in the magnetic resonance imaging examination delay in the development of brain myelinization was determined.

Due to the inadequate mouth opening and agitation during the extraction, general anesthesia was chosen for this particular patient. Clinically observed craniofacial anomalies, severe hypotonia and frequently observed pulmonary infection history lead to the decision of sedation administration.

Inadequate coughing reflex, palatopharyngeal dysfunction and inability to protect the airway because of laryngeal paralysis with poor spasmodic closure of the glottis are lead frequently to aspiration and are not attack secretion sufficiently. In the children with this syndrome because of the minimum esophageal sphincter spasm sometimes gastroesophegeal reflux has been detected and this situation usually results in easy aspiration and precipitates chronic pulmonary diseases.[16] So the girl is planned to take into the operation after 6 hours fasting. In the literature, because of the intravenous and inhalation anesthetics (benzodiazepine and opioids) which is central nervous system depressant, a short-acting anesthetic agents, ketamine is to be on medication for sedation. Antisialogogue administration as a premedication agent in order to prevent secretion aspiration in patients with Moebius syndrome is reported.[17] In our case, low-dose atropine as an antisialagogue agent was premedicated before intravenosus anesthesia.

In cases with Moebius syndrome; central nervous system issues are a prominent feature. In this syndrome patients have emotionless-face because of paralysis of cranial nerves, dispite mental function and capabilities may be normal. Paralysis of the facial nerves and the consequent absence of facial expression reduce the patient's nonverbal community rendering it difficult to evaluate these patient and assess their pain. In our patient, we relied on changes in physiological parameters (heart rate and blood pressure) to assess her level of analgesia.

Four liter per minute of oxygen for securing the potent difficult airway was given during the operation. The case was stable hemodynamically and not desaturated during 30 minutes of operation time and the operation was carried with ketamine sedation and paracetamol analgesia without given any local anesthetic infiltration. After the operation the girl who was awakened with tactile stimulation in 3 minutes, was discharged with overall statement of good.

In these babies with significant hypotonia and high incidence of aspiration, santral respiratory depression agent, e.g. intravenous and inhalation anesthetics, benzodiazepines and opioids must be avoided. Consequently, sedoanalgesia is an alternative anesthesia method, which must be taken in to consideration in short-term surgical intervention, such as tooth extraction with highly predicted difficult intubation patients.

REFERENCES

1. Moebius PJ. On congenital bilateral abducens-facial paralysis. Munch Med Wochenschr 1888;35:91-94.
2. Peleg D, Nelson GM, Williamson RA, et al. Expanded Moebius syndrome. Pediatr Neurol 2001;24:306-09.
3. Fontenelle L, Araujo APQC, Fontana RS. Moebius syndrome: Case report. Arq Neuropsiquiatr 2001;59:812-14.
4. Krajcirik WJ, Azar I, Opperman S, Lear E. Anesthetic management of a patient with Moebius syndrome. Anesth Analg 1985;64:371-72.
5. Kumar D. Moebius syndrome. J Med Genet 1990;27:122-26.
6. Amaya LG, Walker J, Taylor D. Moebius syndrome: A study and report of 18 cases. Binocular Vision Quart 1990;5:119-32.
7. Cronemberger MF, de Castro-Moreira JB, Brunoni D, et al. Ocular and clinical manifestations of Moebius syndrome. J Pediatr Ophthal Strab 2001;38:156-62.
8. Terzis JK, Noah EM. Moebius and Moebius-like patients: Etiology, diagnosis, and treatment options. Clin Plast Surg 2002;29:497- 514.
9. Tuzuner-Oncul AM, Kucukyavuz Z. Prevalance and prediction of difficult intubation in maxillofacial surgery patients. J Oral Maxllofacial Surg 2008;66 (8):1652-58.
10. Ames WA, Shichor TM, Speakman M, Zuker RM, McCaul C. Anesthetic management of children with Moebius sequence. Can J Anaesth 2005;52(8):837-44.
11. Towfighi J, Marks K, Palmer E, et al. Moebius syndrome: Neuropathologic observations. Acta Neuropathol Suppl 1979;48:11-17.
12. Kremer H, Kuyt LP, van den Helm B, et al. Localization of a gene for Moebius syndrome at chromosome 3q by linkage analysis in a Dutch family. Hum Mol Genet 1996;5:1367-71.
13. Slee JJ, Smart RD, Viljoen DL, et al. Deletion of chromosome 13 in Moebius syndrome. J Med Genet 1991;28:413-14.
14. Verzijl HTFM, van der Zwaag B, Cruysberg JRM, Padberg GW. Moebius syndrome redefined: A syndrome of rhombencephalic maldevelopment. Neurology 2003;61:327-33.
15. Hobaika ABS, Neves BS, Fernandes ML, Guedes VC. Anesthesia in a patient with Moebius sequence. Case report Rev Bras Anestesiol 2009;59(3):341-43.
16. Teraz JK, Noah EM. Dynamic restoration in Moebius and Moebius-like patients. Plast Reconstr Surg 2003;111:40-55.
17. Ferguson S. Moebius syndrome: A review of the anaesthetic implications. Paediatr Anaesth 1996;6(1):51-56.

ABOUT THE AUTHORS

Merve N Kadioglu (Corresponding Author)

Resident, Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Ankara University, Besevler, Ankara, Turkey, Phone: +90 505-6846612, Fax: + 90-312-2135474, e-mail: mnkadioglu@gmail.com

Hande Arpaci

Assistant Professor, Department of Oral and Maxillofacial Surgery Faculty of Dentistry, Ankara University, Besevler, Ankara, Turkey

Aysegul M Tuzuner-Oncul

Associate Professor, Department of Oral and Maxillofacial Surgery Faculty of Dentistry, Ankara University, Basevler, Ankara, Turkey