Case Report
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Anesthetic Management of Cleft Palate Associated with Meier-Gorlin Syndrome

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Meier-Gorlin syndrome (MGS) is a very rare autosomal recessive primordial dwarfism disorder, characterized by microtia, patellar aplasia/hypoplasia, and a proportionate short stature. Typical facial characteristics during childhood comprise a small mouth with full lips and micro-retrognathia. Presence of cleft palate in these patients is usual condition. Due to associated congenital pulmonary emphysema and sudden cardiac death during anesthesia these patients present major perioperative challenges to anesthesiologist. We report successful anesthetic care in a 4-year-old male child diagnosed with Meier-Gorlin syndrome and admitted for repair of cleft palate. Because of dysmorphic features, we faced difficulties during intubation. Being a multisystem disorder, each patient of Meier-Gorlin syndrome requires meticulous preoperative evaluation and high level of intraoperative and postoperative continuous monitoring regardless of any surgical procedure. Presented case report highlights the significance of aggressive perioperative management in MGS which can result in successful outcome. As for additional features, it is worth to note that in our patient we didn't reveal short stature and patellar anomalies.

Keywords: Meier-Gorlin syndrome, cleft palate, intubation

Introduction

The Meier–Gorlin syndrome (MGS) is a very rare autosomal recessive disorder characterized by severe intrauterine and postnatal growth retardation, microcephaly, bilateral microtia, and aplasia or hypoplasia of the patella [1]. Many cases have primordial dwarfism with substantial prenatal and postnatal growth retardation [2].

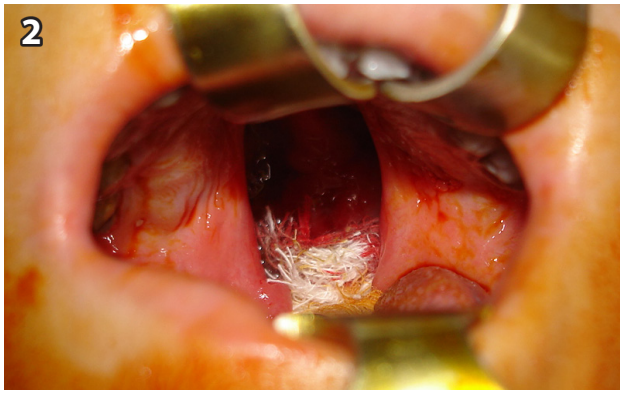
Mutations in five genes from the pre-replication complex (ORC1, ORC4, ORC6, CDT1, and CDC6) were identified in individuals with MGS [3]. MGS is relatively rare. It was defined by Gorlin in 1975, although an earlier case report from 1959 was noted. Since then, additional cases have been reported worldwide [4].

Associated clinical features encompass feeding problems, congenital pulmonary emphysema, mammary hypoplasia in females and urogenital anomalies, such as cryptorchidism and hypoplastic labia minora and majora. Typical facial characteristics during childhood comprise a small mouth with full lips and micro-retrognathia.

Most individuals with MGS have normal intelligence. The acquisition of skills requiring mental and motor coordination (psychomotor development) is normal or borderline normal. Some affected children show delays in attaining developmental milestones.

The diagnosis MGS should be considered in patients with at least two of the three features of the clinical triad of microtia, patellar anomalies, and pre- and postnatal growth retardation. In patients with short stature and/or microtia, the patellae should be assessed with care by ultrasonography before age 6 or radiography thereafter.

Because of multiple system involvement and congenital anomalies the perioperative anesthetic management of these patients is very challenging. We are presenting a report about the perioperative management of case of MGS admitted for repair of cleft palate, which is a usual manifestation of this



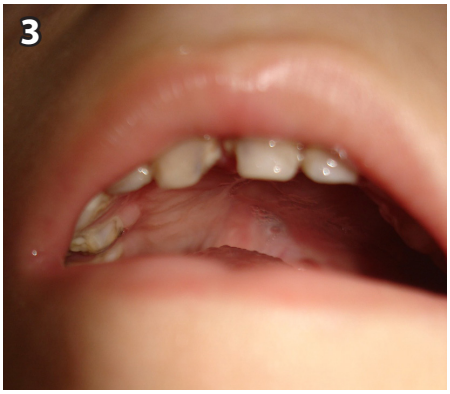


Figure 1.
General view of the patient.

Figure 2.
Intraoral view.

Figure 3.
Postoperative view of repaired palate.

syndrome. As per for our knowledge, after extensive search in medical literature, no such case has been reported in Azerbaijan population and worldwide regarding anesthetic management during cleft palate repair in MGS.

Case report

A 4-year-old boy weighing 10 kg with MGS was referred to our hospital for cleft palate repair. He was diagnosed with MGS by pediatric neurologist at the age of 3. The genetic analysis had shown anomalies in ORC6 gene located in 16th chromosome. The main complaints of his parents were the inability of child to eat solid food and unclear voice. Physical examination revealed characteristic dysmorphic facial features, small mouth, incomplete cleft palate, underdeveloped lower jaw, full lips and a narrow nose with a high nasal bridge (Figure 1, 2). Surprisingly our patient didn't have any patellar anomalies and his height was in normal range. There was no history of seizures, thyroid dysfunction and any surgical procedure in the past. His vital signs were found to be within normal limits for his age. Heart rate was 120/minute and blood pressure 90/40 mmHg, respiratory rate 24/minute, and SpO_2 99% on pulse oxymetry. His biochemical parameters, including specifically investigated serum calcium and thyroid function tests were within normal limits. Chest radiograph, electrocardiogram and echocardiography results were normal as well. His Mallampati score was 4, the thyromental distance was grade 3 and his inter incisor distance was one finger breadth only. Potential risk during anesthesia and surgery was explained to parents in detail and written high risk consent was obtained.

The child was kept nil by mouth for six hours and the morning dose of 10 mg ranitidine tablet was given with sip of water. An intravenous access was taken with 22 G angiocath after applying local anesthetic cream (Emla) and the patient was sedated with oral dose of midazolam 0.5 mg/kg. Half an hour before induction, bolus of antibiotic was administered slowly intravenously. Pre-oxygenation and continuous monitoring with electrocardiography, noninvasive blood pressure, SpO2, peripheral O2 saturation and end tidal CO2 started. After 5 minutes of preoxygenation patient was induced with propofol 2 mg/kg, fentanyl 2 μg/kg and atracurium 0.5 mg/Kg. During laryngoscopy the visualization of vocal cords was difficult due to characteristic facial dysmorphic features; his Cormack and Lehane grade was strong 4. After three unsuccessful attempts with McCoy laryngoscope and with the fiberoptic bronchoscope, an emergency tracheostomy was performed and intubation was done with 3.5 inch cuffed tube. The ventilation delivered in intermittent positive pressure ventilation (IPPV) mode with appropriate parameters.

Cleft palate defect was closed by correcting position of muscles of soft palate and reconstruction of muscle sling. Anesthesia maintained on oxygen, sevoflurane and intermittent doses of atracurium. During intraoperative phase there was one episode of tachycardia which was settled with additional dose of fentanyl (5 microgram) and deepening plane of anesthesia with sevofluran. Vital and hemodynamic parameters were stable and continuously monitored during intraoperative period and patient did not show signs of awareness. Surgery lasted for two hours and after confirming haemostasis, throat packs were removed. After gaining adequate power, patient was extubated. Tracheostomy tube was left, oxygenation continued and patient observed for 15 minutes in operation theatre. With continuous monitoring patient was shifted to surgery unit where he was observed for 24 hours and pain was managed with intravenous paracetamol 30 mg/kg. On second post-op day his tracheostomy tube was removed and on third post-op day he was discharged without any adverse event (Figure 3).

Discussion

Ear-patella-short stature or Meier-Gorlin syndrome is an extremely rare condition, with less than 50 cases reported in the literature. It is an association of malformations which typically include bilateral microtia, absent patellae, short stature, poor weight gain, and characteristic facial features such as high forehead, micrognathia with full lips and small mouth, and accentuated nasolabial folds. In our case, whose diagnosis was proved by genetic analysis, there were no patellar anomalies as well as short stature.

Although cases of Meier-Gorlin syndrome reported since 1959, we could not find any report concerning details of general anesthesia management in these patients. In our case, we confront with some difficulties during perioperative period. These included Mallampati score 4, grade 3 thyromental distance and interincisor distance one finger only. During direct laryngoscopy, his Cormack and Lehane grade was determined to be strong 4. These findings can be partially explained as general tendency for malformations including those in orofacial area, as for instance underdeveloped mandible, temporomandibular joints, tilted larynx and overhanging base of the tongue. Taking all mentioned above into an account the anesthesiology and the surgery teams should be ready for difficult intubation. The prolonged nature of surgery and prone position required mean that the patient would require good relaxation, adequate analgesia and also sufficient oxygenation to tide over the preoperative phase. Therefore, facial or laryngeal mask hardly can be a choice for such cases. If intubation fails after three attempts, then alternative ways of ventilation i.e. jet-ventilation, cricothyrotomy or tracheostomy must be strongly suggested. It is worth to note that operations performed in oral cavity impose particular challenge on respiratory ways, so from alternative ways of intubation, the tracheostomy seems to be the most appropriate.

According to our experience, the patients with Meier-Gorlin syndrome should be investigated before surgery for anomalies in airway, so that both anesthetist and surgeon could have a plan and be ready for emergency situations. Surgical correction of cleft palate in such patients demands very high anesthetic and surgical skills as both share common airway. Being a multisystem disorder, each patient of MGS requires meticulous preoperative evaluation and high level of intraoperative and postoperative continuous monitoring regardless of any surgical procedure. Presented case report highlights the significance of aggressive perioperative management which can result in successful outcome in patients with MGS.

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