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Case Report

Spinal Anaesthesia for Inguinal Herniotomy in an Infant with Mucopolysaccharidosis type III with skeletal dwarfism: A Case Report

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Abstract

Mucopolysaccharidosis (MPS) is an inherited lysosomal storage disorder where glycosaminoglycans accumulate in various connective tissues, resulting into a myriad of organ system abnormalities. One of the most challenging aspect of their anaesthesia management is airway control, as these are almost always deemed difficult airway subjects. Anaesthetising neonates or small infants with MPS proves to be an even more critical scenario due to their added airway and respiratory physiological characteristics. Conducting surgeries on such patients under sole regional/ neuraxial anaesthesia can be an extremely efficacious and safe approach, provided the difficulties in their neuraxial anatomy are kept in mind, and thereof, managed accordingly. However, sole neuraxial anaesthesia in small children with MPS is scantly reported in existing literature and hence needs further reporting and study.

Keywords: Mucopolysaccharidoses, Mucopolysaccharidosis III, Obstructive Sleep Apneas, Dwarfism, Airway management

Introduction

Mucopolysaccharidosis Type III, or Sanfilippo syndrome, is a prevalent autosomal recessive lysosomal storage disorder resulting from impaired heparin sulfate metabolism (reported prevalence: 1:53,000 - 1:360,000). Characterized by four enzyme defects (MPS IIIA-D), MPS IIIA carries the gravest prognosis. Clinically, children with MPS III initially show mild developmental regression, progressing after age two to severe behavioural disorders, intellectual deterioration leading to dementia, potential sleep-disordered breathing, and mild facial dysmorphism. Around ten years, progressive neurological decline, motor regression, spasticity, communication problems, seizures and dysphagia develops [1]. Systemic involvement, including cardiac and skeletal abnormalities, has been reported. While typical survival is around two decades, it can extend to 30-40 years for types B-D [1]. Anesthetic management in these children is challenging due to difficult intubation from airway abnormalities and unstable necks, associated cardiorespiratory disease, and obstructive sleep apnea, with difficult intubation rates reported between 28-44% [2]. Regional anesthesia is often considered safer. This study presents the anesthetic management of a 6-month-old male infant with MPSIII undergoing bilateral inguinal herniotomy under spinal anesthesia with mild sedation.

Report of the case

A 6 month old male baby with diagnosed MPS III, was posted for emergency bilateral inguinal herniotomy for obstructed bilateral inguinal hernia. The baby was born late-preterm at 36 weeks gestational age, by caesarean section due to oligohydramnios, had a birth weight of 1.7 Kg, cried immediately after birth; thereafter, was

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observed in NICU for three days. The baby had one episode of community acquired pneumonia, at 5 month age, which was managed by antibiotics conservatively. However, it was during this hospitalization, that the patient was evaluated for his syndromic appearance and genetic evaluation was carried out. He had multiple skeletal abnormalities- pectus excavatum, skeletal dysplasia, mesomelic dwarfism, restricted movements of small joints. Echocardiogram was normal and whole exome sequencing confirmed the diagnosis of MPS III. Ophthalmological and otolaryngological examination did not reveal any further abnormalities. During pre- anaesthetic evaluation, we found the following: poorly built for his age- weight 3.14 Kg, height 53 cm, head circumference 41 cm, upper segment: lower segment ratio= 1.74:1; with extremely loose lax skin, coarse facial features, prominent forehead, micrognathia and mild retrognathia (Fig. 1). Spine appeared normal on clinical examination. Severe subcostal retractions, grunting and stridor were noticed, along with bilateral coarse vesicular breath sounds and normal heart sounds on auscultation. All developmental milestones were significantly delayed. Parents also reported snoring and noisy breathing. On examination of his airway, we found no predictor of difficult mask ventilation (COPUR score-5), except his slightly dysmorphic facies with a small mandible; however, with the background of grunting, stridor, subcostal retractions, probability of difficult intubation could not be ruled out. In view of emergent nature of surgery, no further investigations or airway imaging were asked for.

Anaesthesia management-

Preoperatively, standard fasting guidelines were followed, postoperative ICU bed ensured, fully informed, written high-risk parental consent obtained. Parents were explained about the plan of spinal anaesthesia, the possibility of requirement of general anaesthesia if the former is difficult, and risks associated with general anaesthesia in this baby including consent for emergency tracheostomy.

Anaesthesia planned was- spinal anaesthesia, with backup of general anaesthesia using supraglottic airway device.

On the day of surgery, OR was appropriately prepared following standard protocol, along with difficult airway cart including video laryngoscope and fibreoptic bronchoscope, warming mattresses, microdrip sets, soft jelly pads and cotton rolls. Inside OR, standard ASA monitors were attached, IV cannula, 22G which was already in situ on the right hand dorsum, was checked for patency and 0.5 mg midazolam was administered IV. Titrated, divided doses of IV Fentanyl (total 5 µg- two doses of 2.5 µg each) and Ketamine (total 5 mg- two doses of 2.5 mg each) were administered, to achieve mild sedation; just enough to ensure that the patient was comfortably asleep but was waking up with painless stimulus like touch. Ease of mask ventilation was checked by gentle bag- mask ventilation, using appropriately sized facemask and ambubag, and then discontinued. Thereafter, O_2 was supplemented by simple facemask at 4-6 liters/minute with $EtCO_2$ monitoring using side-stream capnometry, which confirmed that patient breathed spontaneously. The patient was then positioned in left lateral decubitus, taking care of the neck and all bony prominences. Subarachnoid block was given using 26 G Quincke's paediatric spinal needle, using the midline approach in L5-S1 intervertebral space, and after confirming free flow of CSF, 0.6 ml of 0.5% heavy Bupivacaine (1 mg/Kg actual body weight) was injected slowly. The patient was then positioned supine and loss of sensation to pain (pin prick) was confirmed till T10 dermatomal level, before proceeding for surgery. Vital parameters were stable throughout intraoperative period, total 50 ml of RL was administered and bilateral inguinal herniotomy was completed in 1 hour 15 minutes. The baby woke up fully active at the end of surgery, with slight stimulus. He was monitored for 30 minutes in the PACU, and as per protocol, shifted to general surgical ward once his modified Bromage score was 0 (full flexion of knees and feet) and had passed urine.

Postoperatively, oral feeds were started and patient received paracetamol 15 mg/Kg IV three times a day for the subsequent two days. It followed an uneventful postoperative

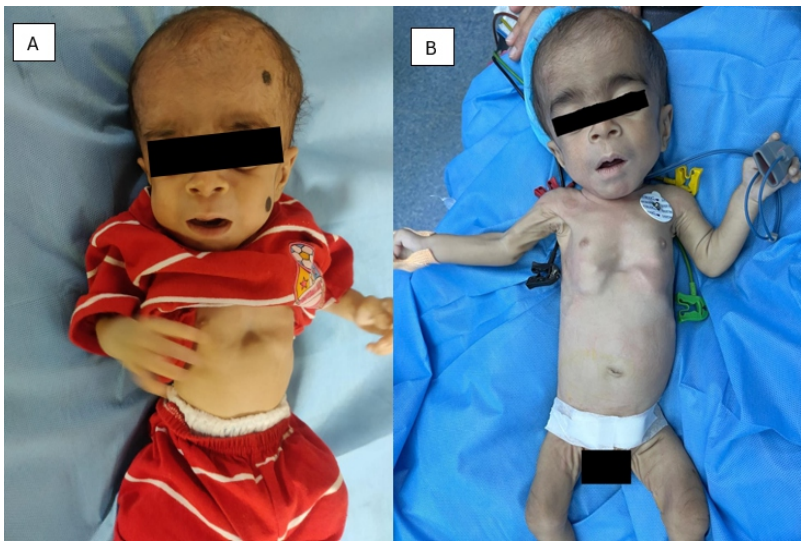

Figure 1: Photograph of the patient during pre- anaesthetic examination (A) and at the end of surgery, breathing spontaneously, subcostal retractions evident (B)

course. On the third postoperative day, he was discharged home.

Discussion

Patients with MPS have multiple comorbidities, commonly requiring surgical interventions. It is associated with unique phenotypic facial and airway characteristics, making perioperative airway management challenging. The overall incidence of difficult mask ventilation in MPS has been reported as 7-14% [3, 4]; and the incidence of difficult tracheal intubation ranges from 24-48% [3, 4, 5]. As such, most of the recent literature suggests, video laryngoscope is being increasingly preferred as the primary airway management plan in these patients by most anaesthesiologists [6]. Not rarely, failed tracheal intubations requiring emergency tracheostomy have also been reported [3, 5, 7]. However, compared to all other types, MPS III poses the least amount of airway challenges for the anaesthesiologists [2, 8].

Besides various airway issues, patients with MPS have other comorbidities that may affect ventilation. They can have restrictive or obstructive lung disease, recurrent lung infections, and obstructive sleep apnea [5]. Propensities for bronchospasm and oxyhemoglobin desaturation must also be kept in mind in these children. Further, skeletal dysplasia such as atlantoaxial instability, spinal cord compression, limited neck mobility, pectus carinatum, and scoliosis is not uncommon [9].

Multiple case reports have documented failure of epidural anaesthesia in MPS patients, owing to deposition of mucopolysaccharides in the general epidural space as well as sheath of nerve fibres [10]. Such problems are obviously not encountered during spinal anaesthesia However, older children with intellectual disabilities may not always comply for spinal anaesthesia in awake condition.

The present case describes a preterm infant with snoring history and stridor, with LRTI, and multiple chest wall skeletal deformities, undergoing emergency bilateral inguinal herniotomy. Being late preterm, postoperative apnoea possibilities should be kept in mind while providing anaesthesia, more so, as history is also suggestive of sleep disordered breathing. Because of the chest wall deformities and existing lung infection, ventilatory capacity of lungs are anticipated to be compromised. Although MPS III rarely presents with significant airway difficulties, since our patient presented with inspiratory stridor, grunting and subcostal retractions, possibility of probable difficult intubation cannot be ruled out in this case. General anaesthesia (GA) would have to be given extremely cautiously, as all these factors might contribute to multiple anaesthetic complications. Hence, airway handling and GA has been completely avoided in our case and surgery was done under spinal anaesthesia after mildly sedating the baby to help in positioning and ensuring a quiet OT atmosphere. However, preparation for general anaesthesia with difficult airway cart was done, in case of failure of neuraxial anaesthesia.

Another concern for our patient was that, he had skeletal dwarfism. These patients are prone to apnoea and sleep disordered breathing, narrow airways [11], which makes them ideal candidates for sole regional anaesthesia, wherever applicable. This is also relevant in calculating drug and IV fluid doses. We have used 50th percentile weight for his height (approximately 5 Kg) for dosing of IV drugs and fluids; in accordance to the protocol followed by paediatricians in our institute for patients with achondroplasia. However, for spinal Bupivacaine dose, his actual body weight (3 Kg) was used, as per existing literature in anaesthesia [12].

To conclude, this case presents the unique challenges of anaesthetizing an infant with MPS along with skeletal dwarfism, posted for an emergency surgery. MPS poses unique anaesthetic challenges that require knowledge of disease-specific concerns, preoperative optimization, preparation and tailor-made anaesthetic management approach, pertaining to individual needs. A multidisciplinary team, consisting of an experienced paediatric anaesthesiologist, with paediatric tertiary care support is ideal for such management. Most of these patients have difficult airways and altered respiratory physiology; regional and neuraxial anaesthesia is a logical choice, wherever suitable. However, establishing its safety and effective use in MPS patients, especially neonates and young infants, requires further large- scale studies, in order to be taken as

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

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