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Challenges in the anaesthetic management of newborn having anorectal malformation associated with occipital encephalocele and microcephaly- A case report

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Abstract: *ARM is commonly associated with duodenal atresia, tracheoesophageal fistula, vertebral and renal anomalies (VACTERL anomalies) but, here we report a case of 3 day old newborn, presented with ARM, occipital encephalocele (posterior) and microcephaly, posted for emergency high loop sigmoid colostomy. Difficult airway was anticipated, however tracheal intubation accomplished without any aid with Kimberly-Clark microcuff paediatric endotracheal tube. Thus, in this case report we highlight a rare association of ARM, occipital encephalocele (posterior) and microcephaly along with anaesthetic challenges we faced in this case those are worth to share.*

Keywords: *ARM (anorectal malformation), occipital encephalocele, microcephaly, difficult airway.*

I. Introduction

Anorectal malformations (ARM) are the birth defects that occur at fifth to seventh weeks of fetal development where the anus and rectum do not develop properly. The average worldwide incidence is 1 in 5000 live births. In addition, ARM occurs in association with several syndromes, it coexists with esophageal fistula, tracheoesophageal fistula, vertebral and renal anomalies (VACTERL anomalies)[1].

Encephalocele is a herniation of cranial contents through a congenital defect in the cranium. The incidence is 1 in 5000 live births[2]. Anterior encephaloceles are more common in males while seventy percent of posterior or occipital encephaloceles[3] occur in females.

Henceforth, we report a case with uncommon association of ARM, occipital encephalocele (posterior) and microcephaly, even though occipital encephalocele (posterior) is not commonly present in males along with this anticipation of difficult intubation[4,5].

II. Case Report

This case was a 3 day old male newborn delivered at term in a peripheral hospital by normal vaginal delivery. The child was transferred to our setup for tertiary care and surgical management of the anomaly. At birth the APGAR score was 10 and weight was 2500 gm. There was a cystic swelling of size 7x8 cm in craniocaudal and side to side dimensions. This swelling was arising from posterior aspect of head and was extending to the neck which was suggestive of an encephalocoel. The head was flattened and the anterior fontanel was wide open. There was also an associated microcephaly. Because of the associated high anorectal malformation the abdomen was hugely distended. There were associated dilated anterior abdominal veins suggesting raised intraabdominal pressure. Spine appeared normal on palpation. Neonate was active and there was no other obvious congenital anomaly. This was also confirmed on ultrasound and X-ray chest-abdomen.

Owing to the huge abdominal distension and presence of high anomaly a high sigmoid loop colostomy was planned.

Blood investigations were in normal range. Neonate was received in operation room with intravenous catheter and NG tube. Neonate was placed over soft pillow and head in two large rings in a slight neck extended attitude and was stabilized with assistance fingers. Pre medication was done with Inj. Atropine 0.03 mg IV (10 mcg/kg IV), Inj. Ondensetron 0.25 mg IV (0.1 mg/kg IV). Induction was done with Inj.thiopentone sodium 12 mg IV (5 mg/kg IV) along with O_2 + N_2O (50:50) with halothane 0.6 %. laryngoscopy done with miller's straight blade no. 1 and tracheal intubation was done with Kimberly- Clark microcuff paediatric endotracheal tube no. 3.Tracheal intubation could be done in a very single attempt without any aid just because the problems possible were anticipated and we were already prepared for them. Anaesthesia was further maintained with O_2 + N_2O (50:50), halothane 0.6 and Inj. Vecuronium 0.2 mg IV using mechanical ventilator.

Intravenous fluid Isolyte P given according to holliday and segar's[6] '4/2/1' rule. ECG, heart rate and SpO_2 monitoring was done throughout the procedure. At the end of surgery muscle relaxant was reversed with Inj. Neostigmine 0.125 mg IV (0.5 mg/kg) and Inj. Atropine 0.06 mg IV (20 mcg/kg). After extubation, neonate was comfortable and maintaining saturation. Neonate was then shifted to NICU ward for postoperative care.

III. Discussion

Anorectal malformations are defects that occur during the fifth to seventh weeks of fetal development. With these defects, the anus and the rectum do not develop properly. Anorectal malformations affect one in 5,000 babies and are slightly more common in males. The exact cause of anorectal malformations is unknown. In some cases, environmental factors or drug exposure during pregnancy may play a role, but this is still unclear. Approximately 50 percent of babies with anorectal malformations have other coexisting abnormalities. These commonly include: Spinal abnormalities, such as hemivertebra, absent vertebra and tethered spinal cord, Kidney and urinary tract malformations, such as horseshoe kidney and duplication of parts of the urinary tract, Congenital heart defects, Tracheal and esophageal defects and disorders, Limb (particularly forearm) defects, Down syndrome, Hirschsprung disease and duodenal atresia[1]. In this case we noted anorectal malformation in association with posterior encephalocoel and microcephaly, which could be the rare association and not yet reported in papers.

Airway management[8,9,10] in pediatric patients with posterior encephalocoel poses many challenges to the anesthesiologist. Anesthetic management of this neonate requires carefully attention because the size of posterior encephalocele is too large which caused restriction of head movement[11]. This led to difficulty in positioning for laryngoscopy and in visualizing the glottic opening. We tried to optimize the patient position for adequate exposure of glottis opening and avoided accidental rupture of posterior encephalocoel during intubation. Careful perioperative management allowed us to achieve successful outcome in this case.

Zeynep Baysel Yıldırım,et al[12] also described two different techniques, one in lateral position and another one taking patients head out of the table with the help of assistance. One modified approach is by placing a silicon support, in our case we placed patient supine while head was placed in two large doughtnut shaped head rings and trunk was supported with foldable towels because we felt that this position was technically better in the intubation and fortunately we could achieve intubation without any difficulty in this position.

To intubate the trachea we used Kimberly- Clark microcuff paediatric endotracheal tube no. 3 to avoid accidental extubation perioperatively. The cuff of this PVC tube was made up of polyurethane membrane with sealing pressure of 11 cm of H_2O to avoid trauma to mucous membrane and glottis oedema. To avoid unwanted rupture of posterior occipital encephalocele, neonate was placed on soft cotton pillow with head supported by large two head rings and with assistance finger. Warm cotton rolls were wrapped in all four limbs, chest and head to prevent hypothermia. We didn't use any benzodiazepine and analgesic perioperatively due to early awakening and fast recovery from the anaesthesia. Neonate was followed up for any croup, respiratory distress and any discernible sign's, all went uneventful.

IV. Conclusion

Anorectal malformation and its association with encephalocoel is a challenge for anaesthetist because of difficult anticipated intubation. A proper preoperative planning can help us in managing these cases.

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