Dandy Walker Syndrome with Giant Occipital Meningocele- A Paediatric Difficult Airway.

Case Report

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ABSTRACT

We present a rare case of Dandy-Walker Syndrome, a congenital brain malformation, accompanied by a giant occipital meningocele. The case highlights the critical role of decision-making during difficult airway management, particularly in pediatric patients. The patient, an 8-month-old female child weighing 5 kg, was brought to our outpatient department with a noticeable giant occipital meningocele. The child was shifted to the operation theatre. Two attempts of direct laryngoscopy failed to visualize even the epiglottis, indicating a challenging airway. To manage the airway, we decided to consider the supraglottic airway as rescue method of ventilation. We used an adult fiberoptic bronchoscope to visualize the glottic opening through the supraglottic airway. Additionally, we inserted a central venous catheter guidewire through the side port of the bronchoscope beyond the vocal cords, which served as a conduit to railroad the endotracheal tube. Overall, the case underscores the importance of quick thinking and appropriate decision-making during challenging airway management, especially in rare and complex cases such as this one.

Key Words: Fiberoptic intubation, guide wire, occipital meningocele, paediatric neuroanaesthesia, supraglottic airway.

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INTRODUCTION

Dandy–Walker syndrome is a rare congenital anomaly characterised by hydrocephalus, posterior fossa cyst and absence of the cerebellar vermis with very few cases reported in association with occipital meningocele[1]. This syndrome is often associated with other anomalies including difficult airway. Airway crisis is an ever-present risk in pediatric neuroanesthesia[2]. In crisis situation, we have either the option to successfully manage the airway or risk the life of patient. Prevention and preparation for crisis beforehand is better by laying out a plan and open communication with the team. We are reporting a case of Dandy Walker Syndrome with giant occipital meningocele with difficult airway highlighting the importance of decision making during the difficult airway management.

CASE REPORT

An 8 months old female child, weighing 5 kg reported to our outpatient department with complaints of progressively increasing occipital cystic swelling since birth. The child was born as a result of caesarean section at term of non-consanguineous parents. It was a twin pregnancy. There was no history of any congenital malformation or neural tube defect in the sibling or family. On examination, a giant occipital swelling larger than the child's head extending from occipital protuberance to mid-thoracic level (Figure1) was noted with a positive transillumination test (Figure2). The child had delayed development milestones. Non-contrast computed tomography head demonstrated a large defect (8.4X7.6 cm) in the occipital bone with a large swelling in the occipital region suggestive of occipito-cervical meningocele. Magnetic resonance imaging of the brain revealed a giant cystic CSF intensity swelling lined by dural sac of size 16 x 13 x 9 cm in the occipital region through a large defect in posterior part of occipital BONE (diameter 8cm).The sac was_communicating with fourth ventricle and upper part of spinal canal with moderate communicating hydrocephalus. Mass effect was seen on bilateral cerebellar hemispheres laterally and brain stem and upper cervical spinal cord with dilatation of bilateral lateral ventricles and vermian hypoplasia. A neuroradiological diagnosis of Dandy Walker Syndrome with a giant occipital meningocele was made. During preoperative visit child had limited neck extension with no neurological deficit and cardiac and respiratory system examination was within normal limits. All haematological investigations were within normal limits.

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Fig.1 Showing Large occipital swelling

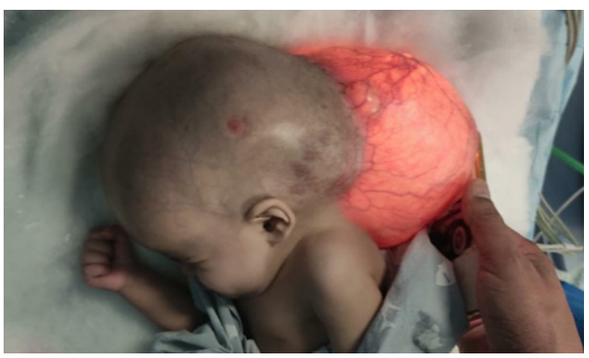

Fig. 2 Showing positive transillumination Test

The child was to be taken for surgery under general anaesthesia in prone position with plan of meningocele repair followed by ventriculoperitoneal shunt surgery. Written informed consent was taken from parents. Preoperatively child was conscious, active, moving all four limbs. Child was shifted inside operation theatre. Intravenous canula was secured beforehand. Standard monitors were attached as per American Society of Anaesthesiologists standard. Vitals were normal. A paediatric difficult airway cart was made available to deal with any possible airway emergency. However, pediatric videolaryngoscope and fiberoptic was not available at our institute.

Child was kept on operation theatre table in right lateral position. Closed circuit was primed with 8% sevoflurane, preoxygenation was done with 100% oxygen with closed fitting mask. Premedication was done with midazolam 0.5 mg and fentanyl 10 ug intravenously and inhalational induction with sevoflurane was done. After checking for mask ventilation, injection succinylcholine 10mg was given and ventilated using sevoflurane, 50% oxygen-air mixture. Attempt at direct laryngoscopy was taken with child in right lateral position. Laryngoscopic blade could not be negotiated due to limited neck extension since the neck mass approximated with the back of the patient and giving no space. Smaller straight laryngoscopic blade could be managed to be inserted however, the epiglottis could not be visualized with it. Subsequently, child was brought to the edge of the table with the head lying out of the table supported by two persons from both sides. Epiglottis could not be visualized in second attempt at laryngoscopy indicating Cormack Lehane grade[4]. All attempts of laryngoscopy were very quick and few bag mask breaths was given in between the successive attempts. No desaturation was noted during this time. After two failed attempts of direct laryngoscopy, immediately supraglottic airway (Classic LMA) of size 2 was inserted and ventilation confirmed. We were able to ventilate with supraglottic airway in supine as well as lateral position. We would have conducted the case with Classic LMA airway in situ, but the duration of surgery was long and meningocele repair had to be carried out in prone position, so definitive airway was required. As the large size of meningocele was causing difficulty in intubation and positioning, meningocele sac was slowly decompressed by draining around 500ml of clear CSF in left lateral position and carefully monitoring patient's vitals. After meningocele sac was decompressed, we gained a little neck extension but it was still not adequate for direct laryngoscopy. We tried inserting pediatric bougie through supraglottic airway blindly, but it failed. Hence, we decided to use adult fiberoptic bronchoscope to visualize vocal cords through supraglottic airway. Adult fiberoptic bronchoscope was inserted through ventilation port in supraglottic airway and glottic opening visualized. An adult central venous catheter guidewire was passed through the side port of the bronchoscope and advanced beyond vocal cords under vision. The fiberoptic bronchoscope was withdrawn by carefully stabilizing the catheter. The endotracheal tube # 4 cuffed was railroaded over the guidewire and passed through supraglottic airway into the trachea. Endotracheal tube was pushed to the maximum limit and supraglottic airway was withdrawn slowly. Once the Endotracheal tube was visualized, it was stabilized to prevent accidental extubation during supraglottic airway removal. Guidewire was removed, and ventilation resumed. Ventilation was confirmed by bilateral auscultation and capnography. Endotracheal tube was fixed carefully. Intraoperative course was uneventful with no complication. After the completion of the surgery patient was reversed, endotracheal tube removed after full recovery from muscle relaxants and patient was shifted to pediatric ICU in stable condition.

DISCUSSION

Airway management is a risky business and anaesthetists are familiar with having to make difficult decisions when complex clinical emergencies arise. One of the challenging situation is airway management in neonates and young infants when associated with congenital anomalies of head and neck[3,4]. Decision-making in such critical situations is crucial for patient outcome.

In the present case, patient's head was in fixed flexion deformity and there was no scope of extension due to restriction to the movement of the head and neck by occipital meningocele. Moreover, there was no access to the front of neck for emergency cricothyroidotomy or tracheostomy.Therefore, margin of safety was almost negligible and any mistake could have led to risk to the life of the patient. Two attempts of laryngoscopy first using Macintosh blade and later Miller straight blade, failed to visualize even epiglottis. Chandran P *et al*[5] recommended careful CSF drainage before intubation for giant occipital meningocele to decrease the size of occipital swelling and gain some extension of the neck. In our case, we could achieve some extension of the neck after CSF drainage but it was still not enough for direct laryngoscopy, hence we decided to proceed with fiberoptic bronchoscopy. In our institute, paediatric fiberoptic bronchoscope was not available, hence we decided to visualize glottic opening through supraglottic airway by adult fiberoptic bronchoscope. David T *et al*[6] has described the use of intubation introducers like guide wire through supraglottic airway for paediatric difficult intubation by passing through adult fiberoptic bronchoscope. Latha Rao *et al*[7] also describes the use of intubation introducers through supraglottic airway like angiographic catheter and stiff guide wire for paediatric difficult airway management with availability of only adult fiberoptic bronchoscope.

Therefore, in the situation of difficult laryngoscopy and intubation, we should know our limitation when to stop subsequent attempts at laryngoscopy and intubation and should consider early decision of using alternative methods of intubation.

Here, we could achieve success at intubation by combining two already known methods like CSF aspiration which improved neck extension and using central venous catheter guide wire as introducer through supraglottic airway.

CONCLUSION

Airway management in child with many congenital anomalies is a great challenge even for an experienced anaesthesiologist. Such difficult airway carries a significant risk for serious patient injury and even death. Strategic preoperative planning to deal with failed intubation is self-evident. It is a wise and deliberate act to stop attempts at direct laryngoscopy, announcing 'Failed direct laryngoscopy' timely and choosing alternate method of securing airway.

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CONFLICT OF INTERESTS

There are no conflicts of interest.

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