CASE REPORT

Long-standing meningocele can be a predictor of difficult airway and postoperative hypoventilation: challenge to the anaesthesiologist

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Accepted 28 January 2016

SUMMARY

A 15-year-old female patient presented with severe pain in the abdomen and obstructive uropathy. She underwent Yang-Monti ileovesicostomy under general anaesthesia. She is a known case of long-standing meningocele (MMC) and presented with its potential complications such as difficult airway and restrictive lung disease. We describe the successful anaesthetic management of a case of anticipated difficult airway and postoperative hypoventilation as a sequel of kyphosis due to MMC. Her airway was secured with fibre optic-guided intubation in a semirecumbent position. Postoperative hypoventilation, hypercarbia and respiratory acidosis were managed conservatively, followed by staged weaning in the intensive care unit. Obstruction of the catheterisable continent channel of the neurogenic bladder itself may present with uropathy and urosepsis, which were also taken care of preoperatively.

BACKGROUND

Paralytic meningocele (MMC) can lead to a structural spine anomaly such as kyphosis.¹ Kyphosis can be defined as a posterior angulation of the spine¹ and is often associated with congenital or paralytic MMC.² Long-standing kyphosis can cause rib crowding, reduced lung expansion and decreased lung compliance leading to restrictive lung disease.¹ The abnormal bending of the spine prevents patients from lying down and, as a result, proper positioning during anaesthesia and surgery can be a challenge. Securing an airway may be difficult due to malalignment of the oral, pharyngeal and laryngeal axes. Advanced airway equipment such as a fibre optic bronchoscope is often required. Postoperative hypoventilation due to restrictive lung disease needs to be managed with staged weaning, postoperative physiotherapy and use of a shorter acting opioid or non-opioid analgesics for pain management.³

CASE PRESENTATION

A 15-year-old female patient presented to the emergency department of a tertiary care hospital with severe pain in the abdomen and inability to pass urine for the past 12 h. She was born with a congenital lumbosacral MMC. Her parents did not seek any treatment as she was asymptomatic and attained developmental milestones on time. With age, the congenital MMC kept growing in size and she gradually developed weakness in the lower

limbs with urinary incontinence (neurogenic bladder). At the age of 8 years, she was incapacitated, almost bedridden, unable to lie down in a supine posture because of her MMC and completely paraplegic. Her parents brought her to the paediatric surgery department where the MMC was repaired along with bladder augmentation by a stomach patch for her rudimentary bladder. Five months later, a tubular structure resembling urethra was reconstructed with appendix and was inserted inside the augmented bladder mucosa to provide her continence (Mitrofanoff appendicovesicostomy). Reviewing her history, our paediatric surgery registrar opined that the reconstructed urethra may be stenosed, kinked or obstructed at the bladder neck causing acute obstruction. Emergency suprapubic cystostomy was done and the patient was posted for Yang-Monti ileovesicostomy for the next day.

Preoperative airway examination in the sitting posture revealed Mallampati class I, normal thyromental distance and adequate neck movements. The patient could not lie down in the supine position for clinical examination (figure 1). On careful evaluation, we found a forward bending of the spine at the thoracic level, which prevented her from lying supine. Chest X-ray (posteroanterior view) showed moderate kyphosis, bilateral rib crowding and reduced volume of thoracic cavity (figure 2). Pulmonary function test revealed preoperative forced expiratory volume in first second (FEV1) as 45% of predicted value and vital capacity (VC) as 48% of predicted value. Echocardiography was normal apart from mild pulmonary hypertension (pulmonary artery pressure 28 mm Hg). Effort tolerance could not be assessed as she was bedridden. The kidney function test was normal and there were no signs of urosepsis such as fever, hypothermia, leucocytosis, leucopenia, tachycardia, tachypnoea or other signs of organ hypoperfusion.

On the operation table, two pillows were supported behind her shoulder blade to stabilise her in the semirecumbent position. Routine monitors were attached and mask ventilation started. The possibility of difficult positioning preventing proper alignment of the airway cautioned us to keep the difficult airway cart ready. A C-MAC video laryngoscope with D blade was our first choice since it is meant for a difficult airway situation. After laryngoscopy, only the tip of the epiglottis was visible but was not liftable and the tube could not be negotiated below the epiglottis even with a stylet.

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To cite: Sardar A, Khanna P, Singh A, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-214456

Figure 1 Patient cannot lie down in supine position.

Without wasting time, fibre optic intubation was tried through an endomask since the patient kept desaturating intermittently during airway manipulation. Fibre optic bronchoscope was difficult to negotiate as tongue fall back was aggravated due to gravity in semi recumbent positioned. Finally, the tongue was pulled out of the mouth with a gauge piece by an assistant and we were successful in securing the airway.

Anaesthesia was maintained with 50% oxygen in air, isoflurane and boluses of atracurium. Fentanyl boluses and paracetamol were given intravenously for pain relief. Throughout the surgery, the airway pressure was high. We could ventilate with adequate tidal volume at a peak pressure of 30 mm Hg. Surgery was uneventful and the obstruction was relieved. Blood loss was minimal. While recovering from anaesthesia, she had poor spontaneous efforts and inadequate tidal volume. There was rapid shallow breathing. Normothermia was ensured and the possibility of residual effect of the muscle relaxant was excluded by a train-of-four count of 0.96. Arterial blood gas was done which showed pO_2 75.67 mm Hg, pCO_2 65.32 mm Hg and pH 7.25. A decision was taken not to extubate the patient and she was shifted to the paediatric surgery intensive care unit (ICU) for elective ventilation.

OUTCOME AND FOLLOW-UP

In the paediatric ICU, the patient was put on synchronised intermittent mandatory ventilation (SIMV) mode. For pain

Figure 2 Chest X-ray of the patient.

relief, paracetamol and fentanyl boluses were given when the visual analogue score was more than 3. Rigorous chest physiotherapy was started. Staged weaning was also started. The patient was extubated on postoperative day 3, discharged on postoperative day 10 and regularly being followed up in our spine clinic.

DISCUSSION

Kyphosis can be an isolated lesion or coexist with scoliosis.¹ It may occur in long-standing congenital MMC with an incidence around 12–20%.² Severe kyphosis due to MMC may be paralytic or congenital. The congenital variety is less common but difficult to manage.⁴ Our patient did not have any bony spinal deformity at birth. In our patient, the MMC increased in size with age and pressed upon the spinal cord leading to paraplegia and development of kyphosis. It has been hypothesised that muscular instability with lack of dorsal bony structures lead to the development of kyphosis in paralytic MMC.⁵ Kyphosis can cause crowding of ribs, reduced chest wall compliance and decreased lung volume leading to restrictive lung disease.^{1–6} Pulmonary function testing (PFT) in such cases reveals decreased forced VC (FVC) and FEV1 with maintained FEV1/FVC ratio.⁷ In spite of all, patient maintains near normal respiration by compensating mechanism such as force-length and force-velocity properties of inspiratory muscles, the Hering-Breuer reflex and increased central drive.⁸ These protective mechanisms are lost intraoperatively by general anaesthesia and use of opioids.⁹ Hence, ventilation becomes challenging and the patient may have peri-operative respiratory complications. Preoperative FEV1 or VC <40% is an independent marker for postoperative elective ventilation.⁶

In this case, a preoperative chest X-ray showed rib crowding and reduced lung volume. Pulmonary function test and echocardiography were ordered as there was a high possibility of restrictive lung disease from the chest X-ray. Our patient had moderate restriction in PFT and developed postoperative hypoventilation, atelectasis, acidosis and respiratory failure. Echocardiography was done to rule out right heart failure and pulmonary artery hypertension. The patient was managed for restrictive lung disease. She was gradually weaned from the ventilator and extubated on day 3. Rigorous physiotherapy and postextubation incentive spirometry continued. Analgesics with lower respiratory depression potentials such as paracetamol and fentanyl were used for pain relief.

Another unique problem that we encountered in this case was the difficult airway in spite of airway examination turning out to be normal in preoperative evaluation. This was primarily due to the patient's inability to lie down supine because of her thoracic kyphosis. As a result, it was impossible to align her oral, pharyngeal and laryngeal axes in a straight line for intubation. Abnormal thoracic configuration can also alter mediastinal anatomy with narrowing of the thoracic cavity resulting in airway compression.⁹ In our patient, a difficult airway was anticipated and managed appropriately with fibre optic intubation. Fibre optic intubation was complicated by tongue fall back in our case, which is a common problem encountered due to loss of tone of the tongue and can be overcome by pulling out the tongue with forceps or spatula.¹⁰

Our patient had a neurogenic bladder which requires a catheterisable continent channel. Appendicovesicostomy, a widely used technique based on the Mitrofenoff principle, was performed on our patient.¹¹ It can have certain continent-stoma-related complications such as obstruction, kinking, stricture of the conduit causing severe pain, discomfort and features of

obstructive uropathy.¹² Even established renal failure has been reported.¹¹ Obstruction complicated by infection and abdominal abscess can cause systemic toxicity.¹³ Careful preoperative evaluation to rule out obstructive symptoms and assessment of renal function status were performed in our patient to prevent any untoward peri-operative complication.

This case demonstrates peri-operative management of a patient with a difficult airway and restrictive lung disease due to long-standing kyphosis. This is probably the first case of its kind where a peri-operative difficult airway and restrictive lung disease have been correlated with long-standing MMC and managed successfully.

Learning points

- ▶ Long-standing meningocele (MMC) may present with its stigmata kyphosis and its sequel restrictive lung disease and with peri-operative difficult airway.
- ▶ Spinal deformity and its anaesthetic implications may be overlooked in a case of surgically corrected MMC in childhood.
- ▶ A continent catheterisable channel itself has complications which should be taken care of preoperatively to plan proper intraoperative management.
- ▶ Detailed preoperative evaluation and investigations can prevent untoward peri-operative complications in such patients.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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