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Anaesthetic Management in a Patient with Syringomyelia and Arnold Chiari Malformation Type 1 with Unilateral Vocal Cord Palsy

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Abstract: ***Patient with Arnold chiari malformation has often limited range of cervical spine extension and they are challenge to anaesthesiologist. It is important to consider alternative methods for screening the airway, because these patients are at increased risk for difficult tracheal intubation and neurological injury during airway management. We have reported 42 years old male diagnosed as ACM type 1 with syringomyelia, associated with unilateral vocal cord palsy was operated for foramen magnum decompression and duroplasty. Fibreoptic tracheal intubation was used to prevent neurological damage associated with neck movement. We have successfully performed the intubation by using the fibreoptic bronchoscope and his postoperative period was uneventful.***

Keywords: Syringomyelia, spinal cord disease, Arnold chiari malformation type 1, unilateral vocal cord palsy, limited neck extension, difficult intubation, fibreoptic intubation.

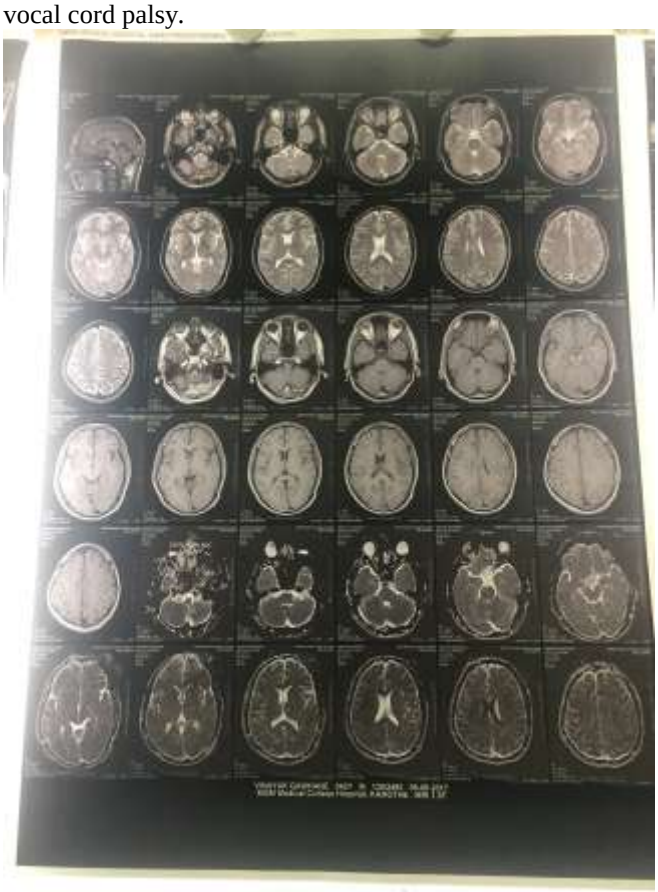
1. Introduction

Arnold chiari malformation (ACM) type 1 is a disorder of certain origin that has been traditionally defined as a tonsillar herniation through the foramen magnum. The anomaly is a leading cause of syringomyelia and occurs in association with bony abnormalities at the cranio-vertebral junction. The natural history is a gradual, stepwise deterioration over many years. ACM type 1 with syringomyelia may manifest with vocal cord palsy, autonomic dysfunction, often asymptomatic and discovered incidentally on brain or cervical spine MRI scan. The main anesthetic concerns are difficult airway management, damage to spinal cord and sensitivity to neuromuscular blocking agents and risk at autonomic dysfunction. We have performed a succesfull management with fibreoptic awake tracheal intubation in a patient with syringomylia with ACM who developed unilateral vocal cord palsy and craniovertibral abnormality with anticipated difficult airway.

2. Case Report

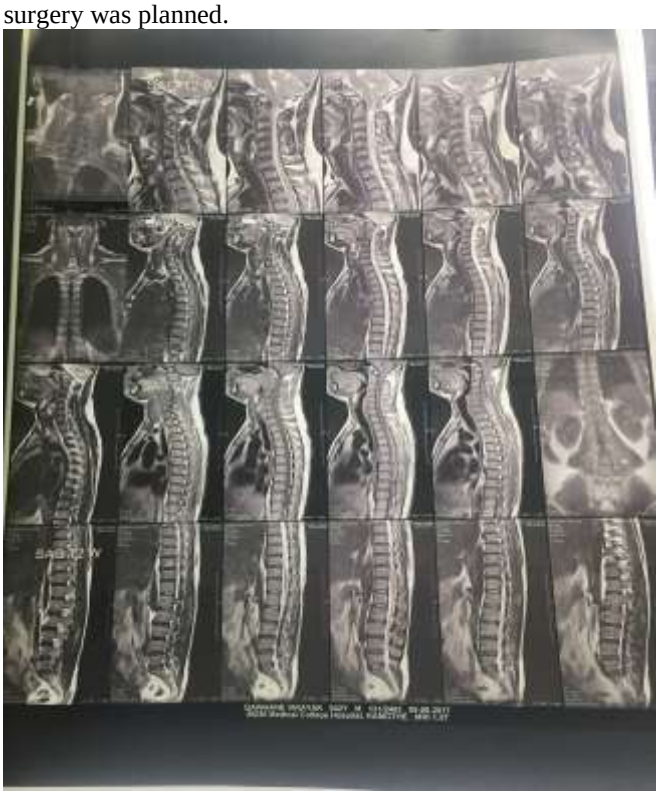
42 year old male patient weighing 70 kg was scheduled for foramen magnum decompression. He presented with hoarseness of voice since 5 days, and episodes of bilateral frontal headache, giddiness and vertigo for 6 months. There were no history of trauma, dysphagia, difficulty in breathing, numbness and weakness of limbs and no any change in bowel and bladder habit.

On physical examination his pulse rate was 120/min, regular, blood pressure was 140/90 mmhg in supine position in right arm. There was no reduced sensation to pain and temperature in any limbs. Routine investigations were within normal limits. Preoperative electrocardiography showed sinus tachycardia and chest X- ray was within normal limit.His airway assessment showed a mallampatti grade 3,thyromental distance was 5 cm short neck and limited extension associated with pain and vertigo. A difficult intubation was anticipated, IDL suggestive of unilateral vocal cord palsy.



MRI brain suggestive of tonsillar ectopic with cevico-medullary kinking causing syringomyelia from C2 to D11 level which is unchanged as compared to prior scan is consistent with Arnold chiari malformation type 1. He was diagnosed as a case of ACM type 1 with syringomyelia and

for that foramen magnum decompression and duroplasty surgery was planned.



It was expected that endotracheal intubation via direct laryngoscopy would not be easy. There was high possibility of neurological damage from extension. Therefore awake fiberoptic bronchoscopy was considered. Difficult airway management cart with rigid bronchoscope and jet ventilation were kept ready. The procedure was explained to the patient and written consent was taken.

Patient was kept fasting for 6 hours for solid and 2 hours for water before OT.Tab Ranitidine 50 mg was given with small sip of water at night before surgery. Patient shifted to OT, Standard monitors (ECG, pulse oxymeter, NIBP)were attached and the baseline vitals were recorded. Inj. Glycopyrolate 0.2 mg i.v. was given. Xylometazoline was instilled in the nostril for vasoconstriction of nasal passage to facilitate passage of fiberoptic bronchoscope(FOB) without mucosal injury.Patient's airway was anesthetized by application of lignocaine 2% jelly in the nostrils, lignocaine viscous 2% gargles, 4 ml of lignocaine 4% nebulization and lignocaine spray 10%. Oxygen was administered via oxygen mask at a rate of 6 L/min. patient was administered 1 mg midazolam and 60 µg fentanyl i.v. for mild sedation. FOB was loaded with 7.0 of ET tube. After explaining to the patient the bronchoscope was inserted through one of the nostrils and advanced towards laryngeal inlet.

Laryngeal and esophageal opening were visualized side by side with laryngeal inlet on the right side. Patient was instructed to take deep breath to facilitate identification of the airway. Then fiberscope was advanced and positioned above carina. Lignocaine 2% was administered via the drug port af FOB as and when required to facilitate the passage of FOB. Endotracheal tube was then threaded over the FOB and the FOB removed. Inj. Propofol 80 mg was administered. Then breathing circuit was attached and the tube placement was confirmed by movement of reservoir bag and capnography. Sevoflurane inhalation was started then inj. Vecuronium 5 mg was administered intravenously. The ET was firmly secured and anesthesia was maintained with O2 in air and sevoflurane, vecuronium and fentanyl. Surgery lasted for 3 hours. Blood loss was replaced with colloids. Intraoperatively patient's vital parameters remained fairly stable. On completion of surgery inj. Ondansetron 6 mg i.v. given to reduce emesis. Residual neuromuscular block was reversed using inj. Neostigmine and glycopyrolate combination at the end of surgery. Patient was extubated when awake, breathing spontaneously and responded to verbal command and shifted to ICU for observation and monitoring. His post operative period was uneventful.

3. Discussion

Type 1 ACM consist of a downward displacement of cerebellar tonsils and the medulla through the foramen magnum and spinal canal to compress the spinal cord, which will causing blockage of spinal canal and stoppage of cerebro-spinal flow.

Syringomyelia is a condition in that a cavity which called a syrinx develops in the spinal cord and filled with cerebro-spinal fluid.

Because of this subarachnoid block increase spinal subarachnoid pulse pressure above the block and produce pressure difference across the obstructed segment of the spinal subarachnoid space, result in syrinx formation and progression. Diagnosis is made through history, neurological examination or brain or cervical spine MRI scan. MRI is choice of diagnostic test for ACM type 1 because it easily show the tonsillar herniation as well as syringomyelia.

Type 1 ACM patient can have
1) Difficulty of airway management.
2) Autonomic dysfunction
3) Sudden increased intracranial pressure.
4) Abnormal sensitivity to neuromuscular blockade.

Patient with these condition can have challenges to the anesthetist during induction, maintenance or emergence of anesthesia and positioning.

1) ACM type 1 is associated with cranio-vertebral bony abnormality including cervical spine fusion at cranio-vertebral junction, which leads to limited range of neck movement. There will be limitation of flexion-extension of neck to prevent compression of neural structure. In this type of cases fibreoptic bronchoscopic intubation instead of standard laryngoscope is recommended in patient with cervical spine abnormality and unstable cervical spine. Our patient had a limited cervical spine mobilization due to bony abnormality and cervical spine fusion at the cranio-vertebral junction. Therefore we have intubated via flexible fibreoptic bronchoscopy from beginning.
2) Autonomic function should also evaluated preoperatively for brainstem involvement. They can be present with postural hypotension, impairment of circulatory reflexes, sinus arrhythmia, postural tachycardia. These patient should closely monitored in the immediate 24 hours after surgery of the risk of sudden cardiac arrest and associated with autonomic dysfunction.
3) sudden increase in intracranial pressure caused by intubation, induction, positioning or extubation leads to spinal cord damage, so anaesthesia technique should be like to avoid increase in ICP. Normocapnia and peak airway pressure should be maintain. Induction agent Propofol we can use safely except ketamine. ABG should be done.
4) These patients have also an increased sensitivity to neuromuscular blocking agents. Succinylcholine should not be used for rapid intubation because it is associated with hyperkalemia, When given in patient with denervated muscle. We can avoid overdosing of non-depolarising neuromuscular blockage by using monitoring of neuromuscular functioning.

4. Conclusion

ACM type 1 and its associated complication and disorder possess anaesthetic risk and safe perioperative management can be achieved by careful attention to the derangements that occur with disease. Outcome will be improved with team management including anaesthesiology, neurology and neurosurgery services.

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