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Anaesthesia Management in Patients with Craniovertebral Junctional Anomalies

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Abstract

Aims: To indentify airway abnormalities, autonomic nervous system dysfunction (ANSD) and postoperative respiratory complications in patients with craniovertebral junction (CVJ) anomalies with Arnold Chiari Malformation(ACM) for safe and better anaesthetic management. *Method:* Forty Three patients with CVJ anomalies with ACM were evaluated and operated under general anaesthesia . *Result* Out of 43 patients of CVJ anomalies with ACM, 29 patients had type I ACM and 14 patients had type II ACM . Vocal cord involvement was seen in three patients in type II. One patient had bilateral vocal cord paralysis (VCP), requiring tracheostomy preoperatively and one had unilateral (left) vocal cord paralysis and another one patient had bilateral vocal cord paresis. Both were planned for awake fiberoptic intubation. Overall vocal cord involvement was 6.9% with 21.42% in type II ACM. No patients had vocal cord involvement in type I ACM. In type II ACM patients, out of eight sleep apnoea patients, three had sleep apnoea with vocal cord involvement and remaining five had sleep apnoea only. Out of those five patients, three patients had difficult intubation. One patient had post operative respiratory failure requiring ventilatory support for two days. Sleep apnoea was seen in 18.6% patients. All patients were belonging to type II ACM with 57.14% incidence. Syringomyelia was found in six patients with four in type I and two in type II ACM. Incidence of syringomyelia in all ACM patients was 13.95 % with 6.89% in type II and 28.5% in type I ACM. Three patients had hydrocephalus in type II ACM. Five patients with cervical collor were planned for awake fiberoptic intubation. *Coclusion:* Preoperative detection of airway abnormality, autonomic nervous system dysfunction , airway security using fiberoptic intubation and post operative airway protection are required for safe anaesthetic management of these patients with CVJ anomalies with ACM.

Keywords: Airway; Anesthesia; CVJ Anomalies; Arnold- Chiari Malformation.

Introduction

The bony abnormalities usually encountered in *craniovertebral junction (*CVJ) anomalies are basilar invagination, occipitalization, and congenital atlantoaxial dislocation [1].

Basilar invagination is the term for a devlopmental anomaly of the CVJ including ACM, syringomyelia, syringobulbia and hydrocephalus [2]. Airway obstruction and restrictive pulmonary disease with cardiovascular manifestations poses a high anaesthetic risk to these patients [3,4].

ACM are structural defects in the cerebellum affecting females more than males. Once it was believed that ACM occured in only one in every thousand birthsbut CT SCAN and MRI suggest increased incidence [5].

There are four types of ACM. Type I and II are common and less severe. Type III and IV are rare but severe. Airway obstruction and restrictive pulmonary disease with cardiovascular manifestations poses a high anaesthetic risk to these patients [3,4].

In type I ACM patients lower part of cerebellum extends into foramen magnum,commonly observed

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in older children and adults. In type II ACM, cerebellum and brain stem extend into foramen magnum. Other disorders found are syringomyelia, hydrocephalus, tathered spinal cord, scoliosis, kyphosis, cranial nerve dysfunction [5]. People may have dizziness , muscle weakness, numbness, vision problem, headache, problem with balance and co-ordination, swallowing difficulties, sleep apnoea, breathing irregularities and weakness of vocal cords [2,5-9].

The aim is to provide safe anaesthesia with detailed knowledge of CVJ anomalies with ACM in association with airway abnormalities, syringomyelia, hydrocephalus , autonomic nervous system dysfunction with proper postoperative management.

Method

A retrospective analysis of 43 patients with CVJ anomalies with ACM was done. All patients were evaluated preoperatively in the form of age at the time of diagnosis, sex, type of ACM, symptoms at initial presentation, physical findings including airway assessment by mallampati grading, interincisor distance, radiographic findings (MRI and CT SCAN) and type of surgery. The results of flexible fiberoptic laryngoscopy, methods of airway management like intubating larybgeal mask airway (ILMA), awake fiberoptic intubation(AFOI), tracheostomy and decompressive procedures were noted.

For premedication sedatives were not used.All type I ACM patients and one type II ACM patient were given standard general anaesthesia with assumption of difficult airway and autonomic nervous system dysfunction.

In operation theatre all routine monitors with central venous pressure (CVP) and invasive blood pressure (IBP) monitoring were used. Intubation was done with IV glyco 0.4 ug/kg +fentanyl (4 ug/kg)+pentothal (5mg/kg)+ atracurium(0.5mg/kg) and proper size flexometalic tube was used for intubation with manual inline stabilization. Proper size Ryle's tube was inserted. For maintainance of anaesthesia 0_2+ N_2O (50:50)+isoflurane+ atracurium infusion were used.

At the end of surgery, after return of spontaneous breathing , patients were reversed with glyco 0.8 ug/kg and neostigmine 0.5 mg/kg intravenously.

All patients were extubated when they were fully awake and postoperatively they were monitored for respiratory depression and consciousness level along with vital data.

Results

Out of total 43 patients age at the time of diagnosis ranged from 12 to 42 years with male to female ratio of 20:23. Total number of type I ACM patients were 29 and type II were 14. No patients had type III and IV ACM. Patients were followed up from preanaesthetic assessment to ICU discharge. Most of the patients were presented with the symptoms of pain, motor weakness sensory loss, craniomegaly, stridor and obstructive sleep apnoea. Less common symptoms were growth retardation, dysphagia, seizure, disequilibrium.

Physical findings were motor weakness, sensory loss, hydrocephalus, airway abnormalities due to vocal cord involvement and presence of cervical collar. Surgical procedures done were foramen magnum decompression and occipitocervical fixation,V-P shunt, transoralodontoidectomy.

Eight patients out of 14 patients of type II ACM had obstructive sleep apnoea . Out of which three patients had vocal cord involved with sleep apnoea and five had sleep apnoea alone. Out of three vocal cord involvement patients, one patients had bilateral vocal cord paralysis with near total obstruction. Tracheostomy was done preoperatively and emergency V-P shunt for hydrocephalus was done. One patients had unilateral (left) vocal cord paralysis and one had bilateral vocal cord paresis. Both were planned for awake fiberoptic intubation and needed post operative tracheostomy. No patient in ACM I had airway abnormality. Out of five sleep apnoea patients in type II ACM three patients had difficult intubation and intubated successfully using makintosh laryngoscope blade in second and third attempts. For all these patients preparations for difficult intubation were kept ready.

Five patients were re-explored with cervical collar in situ. In three patients transoral odontoidectomy was done and posted for posterior fixation in second sitting. One patient came for implant removal and one was posted for revision of vp shunt. All those five patients were planned for awake fiberoptic intubation and one had undergone respiratory failure requiring tracheostomy and ventilatory support pastoperatively for two days. As in all these patients mallampati score were three or four and mouth opening one or two fingers, they were planned for awake fiberoptic intubation. Neck

movements were not assesed in view of CVJ anomaly along with neurological dysfunction.

Out of six patients of syryngomyelia in all ACM patients four were from type I ACM and two from type II ACM patients with no autonomic nervous system dysfunction.

One patient in type II ACM had growth retardation.

Discussion

There are five important concerns that an anesthetist needs to pay attention in patients with CVJ anomalies with ACM

a. Patient positioning
b. Difficult airway
c. Neurological monitoring
d. Blood loss
e. Associated medical ailments including chronic pulmonary disease and cardiac disease.

Patients with potentially unstable necks require induction of anesthesia with minimal or no neck movement to prevent spinal cord damage [12].

ACM type I anomaly is associated with descent of hindbrain structures through foramen magnum. There is caudal displacement of cerebellar tonsils and is largely seen in adult population. It is common and less severe. ACM type II is usually seen in children. It is known as classic Arnold Chiari Malformation. There is extension of cerebellum and brainstem into foramen magnum. ACM Type III and type IV are rare as well as severe abnormalities.

Other conditions associated with ACM are hydrocephalus, syringomyelia, scoliosis, kyphosis, tethered spinal cord, some connective tissue disorders and spinabifida [5,8].

MRI is most often used to diagnose chiari malformations as it is sensitive, specific and non-invasive. X-ray cervical spine and CT SCAN can also be used but not much beneficial compared to MRI.

Several surgeries like foramen magnum decompression, cervical laminectomy, duraplasty and VP shunt procedures are used to relieve pressure on the brain and spinal cord and re-establish normal fluid circulation through and around the area [3,11].

Patients usually become symptomatic in the third or fourth decade of life. Headache and neck pain are found in 15 to 75% of patients. Progressive neurological dysfunction including lower cranial nerve deficits are also seen. In type I ACM patients scoliosis occurs in 25% and syringomyelia in 50 to 76% patients.Other airway abnormalities are vocal cord involvement, sleep apnoea and aspiration also [6].

ACM type II and associated hydrocephalus will have symptoms of brain stem dysfunction, stridor, dysphagia, sleep apnoea and cyanosis also. Manifestations of vocal cord impairment may range from unilateral or bilateral vocal cord paralysis to paresis also. Abductor or adductor palsy can occur [2,3].

In our study incidence of vocal cord impairment was 21.42% in type II and no vocal cord involvement was there in type I ACM. Preoperative flexible fiber optic laryngoscopy is advised in all patients with ACM to identify any pre-existing vocal cord impairment to accurately document the incidence of airway abnormality. Even after decompression surgery vocal cord impairment may not recover and tracheostomy may be needed to secure the airway.

In our ACM II patients, one patient needed tracheostomy preoperatively and two patients postoperatively. We found sleep apnoea in eight type II patients. Overall incidence is 18.6%, out of which type II is 57.14% and type I is nil. Three ACM II patients had difficulty in swallowing also.

Movements at the atlantoaxial joint may be prevented by the use of fibreoptic, awake intubation and by placing the cervical collar in a position during intubation and positioning.

A video laryngoscope can be used as an alternative intubation technique. Another option is represented by insertion of an intubating laryngeal mask airway and supraglottis devices (I gel) that allow ventilation and oxygenation during endotracheal intubation attempts [14,16,17].

In our study awake fiberoptic intubation was done in 64% of type II ACM patients.In one study it was stated that Airtraq laryngoscope was used successfully for tracheal intubation for difficult intubation in a parturient with ACM I with syringomyelia as tracheal intubation assisted by gum elastic bougie had failed after two attempts [10].

Syringomyelia is an unusual neurological condition characterised by presence of fluid filled cavity or syrinx within the spinal cord with incidence of 8.4 per 1,00,000 and more seen in male than in women in the 3rd or 4th decade of life [2].

We observed four patients of syringomyelia in type I ACM and two patients in type II ACM. No

patient had autonomic nervous system dysfunction or cranial nerve involvement.

Following problems may arise during anaesthetic management of these patients

- Increased intracranial pressure resulting in damage to spinal cord could be managed by using opioids intraoperatively.
- Sudden cardiac or respiratory arrest due to abnormality in autonomic nervous system. There may be tachyarrythmia and fluctuating blood pressure.
- Airway abnormality resulting in difficult intubation.
- Difficult venous access due to skin lesion.
- Careful positioning of patient is required due to joint deformities.
- Abnormal response to non-depolarising muscle relaxant which can be controlled by use of monitoring and pharmacological reversal.
- Ventilation-perfusion abnormality when there is respiratory muscle involvement.

Evaluation of autonomic functions should be done in patients with significant brain stem involvement for better management during surgery. We should monitor CVP for fluids status and IBP for continuous blood pressure monitoring. Close monitoring iss required in the intermediate 12-24 hours postoperative period. For foramen magnum decompression prone position is used. We should have a plan to reverse the position to supine if there is hemodynamic derangement.

Lokesh Kumar Anand et al [4] found hypotension and tachycardia during foramen magnum decompression. Then supine position was immediately resumed and I.V fluids and phenylephrine were used for management. No patient in our study had ANS dysfunction.

Postoperatively respiratory depression may occur and tracheal extubation should be done when patient is fully awake. Close monitoring of respiratory drive and conscious state needed as indirect indicator of brain stem compression.

In our study one patient was reintubated in postoperative period after 12 hours of shifting and ventilatory support was needed for two days. That patient was 12 years old male with 25 kg weight and MRI findings were CV anomaly in the form parieto occipitalization of C_1 and atlantoaxia subluxation with myelopathy changes and ACM type I. Foramen magnum decompression with C_1-C_2 fixation with bone grafting was done in that patient. When patient had cranial nerve dysfunction and exploration of 4^{th} ventricle is planned, spontaneous ventilation with pressure support for 12-24 hours postoperatively may help in airway protection and respiratory drive monitoring in post operative period [15]. However keeping ET tube in situ requires sedation, this may hamper evaluation of neurological status. So keep ready difficult airway cart to an unusual respiratory compromise. Postoperative pateient requires steroid prophylaxis to reduce edema, bi level positive airway pressure, continuous positive airway pressure in pateint with upper airway obstruction and continuous monitoring of cardiac and respiratory function with physiotherapy for several days after the definitive surgery. IIn in CVJ anomaly patients there may be associated weakness and dysfunction of respiratory muscles, including the diaphragm. Direct compression of medulla due to bony anomalies results in damage to the respiratory center and lower cranial nerve dysfunction which may cause poor gag and cough reflex resulting in frequent aspiration and pulmonary infection.

In one study S.S. Kale found pediatric bony CVS anomalies with five patients of down syndrome and two patients of Klippel-Fleil syndrome [11]. Growth hormone deficiency in children is believed to be a physiological mechanism for inadequate development of posterior fossa with resultant tonsillar herniation [8]. We found a patient with growth retardation in our study.

We had four patients of hydrocephalus. In type I ACM, its incidence is 4-18% [8].

There is concern of difficult airway security, maintenance of immobilization in cervical spine during airway management and positioning apart from concern related to hydrocephalus. Premedication should be used carefully as it may exacerbate or mask signs of neurological dysfunctions. An anxious patient may have rise in ICP during induction and exaggerated hemodynamic changes due to surgical manipulation near medulla in patients with hydrocephalus [13].

Conclusion

A detailed knowledge of abnormalities associated with CVJ anomaliy with ACM like difficult airway abnormality, autonomic dysfunction helps us to recognise and manage the disease appropriately. For all ACM patients flexible fibeeroptic laryngoscope is needed to document vocal cord involvement in airway evaluation.

A safe anaesthetic management should focus on securing the airway with minimum manipulation of neck, preferably awake fiberoptic intubation, hemodynamic changes during posteerriioorr fixation due to prone position, handling of CVJ, autonomic nervous system dysfunction and blood loss during odontoidectomy necessitating the need for CVP and IBP monitoring. Postoperatively ET tube should be kept in situ for 24 hours to prevent complication of reintubation due to pharyngeal oedema with fixed cervical spine and changes of respiratory depression. Tracheostomy may be necessary to secure the airway of patients with VCP, sleep apnoea and to prevent aspiration.

Early recognition and management is life saving in these abnormalities.

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