Clinical Reports

Chiari type I malformation and postoperative respiratory failure

Daryl L. Williams MBBS,
Hamed Umedaly MD FRCPC,
I. Lynn Martin MD FRCPC,
Anthony Boulton MD FRCPC

Purpose: To present a case of respiratory failure following suboccipital craniectomy for Chiari type I malformation.

Clinical Features: A 22-yr-old man presented with a two year history of symptoms and signs suggestive of brain-stem compression at the level of the foramen magnum. This was confirmed with magnetic resonance imaging. The procedure of suboccipital craniectomy, upper cervical laminectomy and fourth ventricle exploration was performed. Three hours postoperatively the patient experienced episodes of apnea and subsequently became drowsy. Blood gas analysis revealed hypercapnic respiratory failure. Chest X-ray revealed evidence of pulmonary aspiration. The trachea was re-intubated and the lungs ventilated in intensive care for 72 hr. He was discharged home two weeks postoperatively.

Conclusion: Chiari type I malformation is associated with a number of associated anomalies. These patients are at considerable risk of respiratory depression and bulbar dysfunction in the perioperative period. The anesthetic issues are reviewed and discussed.

Objectif : Présenter un cas d'insuffisance respiratoire survenue à la suite d'une craniectomie pour malformation d'Arnold-Chiari de type I.

Éléments cliniques : Un homme de 22 ans présentait, depuis deux ans, des symptômes et des signes apparentés à une compression du tronc cérébral au niveau du trou occipital. Ce qui a été confirmé par un examen de résonance magnétique. On a réalisé une craniectomie sous-occipitale, une laminectomie cervicale supérieure et une exploration du quatrième ventricule. Trois heures après l'opération, le patient a présenté des épisodes d'apnée et est devenu somnolent. La gazométrie a révélé une insuffisance respiratoire hypercapnique. La radiographie a mis en évidence une aspiration pulmonaire. La trachée a été intubée de nouveau et les poumons ventilés en soins intensifs pendant 72 h. Le patient a reçu son congé deux semaines après l'intervention.

Conclusion : La malformation d'Arnold-Chiari de type I est associée à de nombreuses anomalies. Les patients atteints sont très à risque de subir une dépression respiratoire et une dysfonction bulbaire en période postopératoire. Les considérations anesthésiques sont passées en revue et étudiées.

From the Department of Anesthesia, University of British Columbia, Vancouver, British Columbia, Canada.

Address correspondence to: Dr. Lynn Martin, Vancouver General Hospital and Health Sciences Centre, 910 West 10th Avenue, Vancouver, B.C., Canada V5Z 4E3. Phone: 604-875-4575; Fax: 604-875-5344; E-mail: lmartin@vanhosp.bc.ca

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CHIARI type I malformation is characterized by descent of cerebellar tonsils through the foramen magnum.¹ Other neurological and skeletal anomalies are often associated with this malformation. Type I malformation occurs mainly in adults.² Suboccipital craniectomy is the commonest surgical intervention performed for patients with objective neurologic deficits.³ We present a case of delayed postoperative respiratory depression following this procedure.

Case report

A 22-yr-old man presented for posterior fossa craniectomy. The patient had undergone lateral rectus adjustment for strabismus two years previously. This had been performed uneventfully under laryngeal mask general anesthesia. He subsequently developed nasopharyngeal incompetence when swallowing and progressive gait disturbance. A diagnosis of Chiari I malformation was made with magnetic resonance imaging (MRI) which revealed cerebellar tonsils herniated to the level of the second cervical vertebrae, crowding of the foramen magnum, and virtual obliteration of the subarachnoid space. The patient was otherwise well and had excellent functional status as a university student. The patient's weight was 62 kg and height was 168 cm. The blood pressure was 110/80 mmHg supine with no postural hypotension. The resting heart rate was 65 beats·min⁻¹ with normal heart rate variability in response to respiration and the Valsalva manoeuvre. Cervical spine mobility was normal and airway examination was unremarkable. Neurological examination revealed diplopia (VI nerve palsies), soft palate elevation to the left (IX nerve palsy), gait ataxia and spasticity in all limbs. There was no evidence of facial weakness (VII nerve), hearing impairment (VIII nerve), voice disturbance or impaired gag reflex (X nerve), or abnormal tongue mobility (XII nerve).

The patient was premedicated with 150 mg ranitidine and 10 mg metoclopramide *po* prior to surgery. Intraoperatively, a 16G intravenous cannula was inserted and routine monitoring (ECG, non-invasive blood pressure, oximetry and capnography) was applied. Additional monitoring included a peripheral nerve stimulator, intra-arterial and central venous pressure monitoring. A modified rapid sequence induction was performed with cricoid pressure. Anesthesia was induced with 200 µg fentanyl, 130 mg propofol and 50 mg rocuronium *iv*. The trachea was intubated with a size 8.5 endotracheal tube (ETT). Anesthesia was maintained with oxygen-nitrous oxide mixture (ratio 1:2) and isoflurane (end tidal concentration <0.7%). A right internal jugular central line was inserted after induction. The patient was positioned prone in a Mayfield head

tongs. Muscle paralysis was maintained with intermittent doses of rocuronium to achieve a post-tetanic count (PTC) 10. Dexamethasone, 10 mg *iv*, was administered to reduce edema.

The surgical procedure of suboccipital craniectomy, cervical laminectomy (C₁ and C₂) and fourth ventricle exploration proceeded uneventfully. There was no evidence of hemodynamic instability, venous air embolus or major blood loss. No further opioids were required during the case. A total dose of 120 mg rocuronium was administered over four hours. At the conclusion of the case, muscle relaxation was reversed with 2.5 mg neostigmine and 0.4 mg glycopyrrolate *iv*. Four twitches were present on train-of-four (TOF) testing and there was no obvious fade on double-burst stimulation. The patient was breathing spontaneously, obeying commands to move all limbs and could maintain sustained headlift. A brisk gag reflex was easily elicited in response to suctioning of the oropharynx. The trachea was extubated when the patient was awake and positioned head-up at 30.

In the post-anesthetic care unit (PACU) the respiratory rate (RR) was 10-16 breaths·min⁻¹ (bpm) and oxygenation was well maintained (SaO₂ 95-96%) breathing 4 l·min⁻¹ O₂ via a face mask. Initial arterial blood gas (ABG) analysis showed normal postoperative ventilation and gas exchange (pH 7.37, PaCO₂ 44 mmHg, PaO₂ 134 mmHg). Chest X-ray revealed normal pleural spaces, normal lung fields and a well placed central line. The patient was alert, oriented and vocalizing normally. Gag reflex was intact. No supplementary analgesics were required. The patient remained awake and alert for the first three hours after surgery. Respiratory rate was 8-15 bpm and oxygenation was well maintained on mask oxygen.

Three hours postoperatively the patient's respiratory rate became extremely irregular (RR 4-8), with apneic spells of up to 20 sec. The patient then became drowsy and systemic oxygenation also deteriorated (SaO₂ 92%) on high flow mask oxygen. Blood gas analysis revealed hypercapnic respiratory failure (pH 7.26, PaCO₂ 70 mmHg, PaO₂ 95 mmHg). Repeat CXR showed a right-sided pulmonary infiltrate suggestive of pulmonary aspiration. The patient's breathing became progressively more periodic and gag reflex became extremely sluggish. Peripheral nerve stimulation revealed no evidence of recurarization. The trachea was re-intubated in PACU for airway protection and to facilitate ventilatory support.

Ventilation and oxygenation were easily maintained after re-intubation (pH 7.37, PaCO₂ 48 mmHg, PaO₂ 167 mmHg, inspired O₂ 40%, PEEP 5 cmH₂O). Urgent non-contrast CT scan showed no evidence of

posterior fossa hematoma. The lungs were ventilated in the intensive care unit. Dexamethasone was continued, a mild diuresis was induced (CVP 5-8 mmHg), and a course of broad-spectrum antibiotics was administered. Respiratory pattern and swallowing improved over the course of 72 hr. The trachea was extubated on the fourth postoperative day, and the patient was closely monitored in ICU for a further three days. He was discharged home two weeks postoperatively.

Discussion

Chiari type I abnormality is one of a group of anomalies associated with descent of hindbrain structures through the foramen magnum. Chiari divided these into four classes in 1891.¹ Type I malformation consists of caudal displacement of the cerebellar tonsils and is largely seen in the adult population. The mean age at onset of symptoms is 25-40 yr.^{4,5} Type I anomaly is frequently associated with craniocervical abnormalities. Our patient had occipitalization (assimilation) of the first cervical vertebra, basilar impression (Figure) and fusion of C₂₋₃ (Klippel-Feil). Kyphoscoliosis, pes cavus and cervical ribs are other bony abnormalities sometimes seen.

Type II anomaly is a more severe abnormality, presenting in infancy and early childhood, whereby the medial structures (cerebellar vermis, brainstem and fourth ventricle) herniate below the foramen magnum. A lumbar myelomeningocele is associated in 95% of cases.⁶ Type III & IV malformations are very severe abnormalities that are exceedingly rare.

The principal aim of surgery in Chiari anomaly is to alleviate local foramen magnum compression and to achieve equilibration of CSF pressures between ventricles and subarachnoid space.³ The most frequently performed procedure is suboccipital craniectomy and C₁₋₃ laminectomy. Fourth ventricle exploration and dural fascial graft interposition between the edges of a dural incision is also often performed as was the case in our patient. We were faced with an interesting dilemma postoperatively because our patient was at considerable risk of brainstem dysfunction resulting in hypercapnic respiratory failure and pulmonary aspiration. We anticipated these complications and chose to extubate the trachea when the patient was awake, nurse him head up, and closely monitor respiratory pattern and level of consciousness as indirect indicators of brainstem dysfunction. Our anesthetic management included thorough preoperative assessment, and our principle aim was to optimise patient outcome in the perioperative period.

A detailed preoperative assessment of Chiari I patients is critical in identifying anesthesia-related

FIGURE Postoperative sagittal MRI Brain demonstrating basilar invagination of the medulla due to a high riding dens (solid white arrow), and herniation of the cerebellar tonsils (solid black arrow). On this T2-weighted image the CSF is white.

perioperative problems. Craniocervical abnormalities are frequently associated with Chiari malformation. Our patient had Klippel-Feil anomaly that was only obvious at the time of surgery. There was no limitation of cervical flexion or atlanto-occipital extension in our patient, but more progressive disease can result in difficult airway management.

Patients may be at risk for pulmonary aspiration due to delayed gastric emptying from autonomic dysfunction and/or impaired deglutition due to impaired upper airway reflexes. Our patient had glossopharyngeal nerve involvement, so we premedicated our patient with a pro-motility agent and histamine type-2 blocker to reduce the risk and/or severity of pulmonary aspiration.

Abnormal vocalization or stridor may be present which can be indicative of vagal nerve involvement. Vocal cord abductor paresis results in extrathoracic obstruction and a characteristic flattening of the inspiratory curve on the flow-volume loop of lung function tests is present. If suspected then spirometry should be performed.⁷

Clinical assessment for autonomic disturbance is mandatory in patients with Chiari I anomaly. Foramen magnum compression may cause intermittent obstruction to CSF outflow resulting in a pressure differential between the cranium and spine.⁴ These abnormal CSF pressure gradients are thought to cause clefts in the floor of the fourth ventricle and cause disruption of the descending tract of the trigeminal nerve, the nucleus

tractus solitarius, and the nucleus ambiguus. Resultant autonomic disturbance is well recognised.⁸ Patients may be asymptomatic but may have subclinical evidence of pathology. Bedside testing should include cardiovascular responses to vagal and positional manoeuvres. Our patient had a resting heart rate in the normal range with normal beat-to-beat variability indicating intact autonomic function. Identification of abnormal autonomic responsiveness is a known risk factor for perioperative hemodynamic instability⁸ and postoperative respiratory depression.

More severe CSF pressure differentials may lead to cystic cavity formation in the spinal cord (syringomyelia). These patients often have impaired respiratory mechanics due to kyphoscoliosis and disordered function of the accessory muscles of respiration. Spirometry should be performed in these cases.

Intraoperatively, we chose "balanced" anesthesia using fentanyl, isoflurane and oxygen-nitrous oxide mixture. We avoided the use of succinylcholine due to the risk of hyperkalemia in a patient with neurological involvement, and instead used a modified rapid sequence induction with high-dose rocuronium.

The commonest postoperative complication is respiratory depression. We chose to extubate the trachea when the patient was awake, and closely monitor respiratory drive and conscious state as an indirect indicators of brainstem compression. Our patient suffered from delayed hypercapnic respiratory failure (Ondine's curse). The etiology of respiratory failure was probably multifactorial. Firstly, central respiratory centre dysfunction may result from either local edema causing compression of the medulla on the clivus or alternatively from vascular spasm causing brainstem ischaemia.⁸ Local edema was more likely in our case because there was marked basilar impression and assimilation of the atlas, and our patient had the fourth ventricle explored intraoperatively. Secondly, our patient had evidence of dysfunction of the ninth cranial nerve, which is the primary afferent neural pathway of the carotid body. Decreased ventilatory response to carbon dioxide,⁹ and insensitivity to hypoxia in association with lower cranial nerve involvement¹⁰ have been described in Chiari malformation. Furthermore CSF pressure fluxes may have partially disrupted the nucleus tractus solitarius which is the primary relay centre for afferents from the aortic and carotid body chemoreceptors.¹¹

We nursed the patient head up and observed him in a fully monitored environment. Our patient had deficient upper airway reflexes and suffered pulmonary aspiration postoperatively. Some, clinicians have advocated elective mechanical ventilation for patients with significant cranial nerve involvement.¹² Certainly, a

lower threshold for maintaining an ETT for airway protection should be considered in patients with: (I) Cranial nerve dysfunction; and when (II) Exploration/decompression of the fourth ventricle is performed. In these cases spontaneous ventilation with pressure support for 12-24 hr postoperatively may achieve both airway protection and respiratory drive monitoring in the postoperative period.

We recommend heightened awareness of problems with airway and ventilatory function in these patients perioperatively. Patients should have: (I) Intensive cardiorespiratory monitoring and observation; (II) An ETT if airway protection or patency is an issue; and (III) Mechanical ventilation if impaired brainstem mediated control is present preoperatively or evident postoperatively.

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