Anesthesia for Cesarean section and posterior fossa craniotomy in a patient with von Hippel-Lindau disease

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Purpose: To describe the care of a pregnant woman with von Hippel-Lindau disease (VHLD) and intracranial mass lesions.

Clinical features: A 30-yr-old primigravida with VHLD at 35 weeks gestation was seen at the obstetric anesthesia clinic because she wished an epidural analgesia during labour. She had a history of headaches and dizziness. Further investigations showed an enlarged cerebellar hemangioblastoma with significant local mass effects. A combined Cesarean section delivery and posterior fossa craniotomy was performed at 37 weeks gestation. A general anesthetic with fentanyl, rocuronium, nitrous oxide, oxygen and isoflurane was given for Cesarean section delivery. After delivery, isoflurane was reduced and propofol infusion at 4–8 $mg \cdot kg^{-1} \cdot hr^{-1}$ was initiated. The patient had an uneventful operative course and recovery.

Conclusions: Patients with VHLD may have worsening of preexisting lesions or develop other lesions during pregnancy. Some asymptomatic lesions can increase the risk for anesthesia complications. These patients need comprehensive assessment before administration of anesthesia.

Objectif : Décrire l'anesthésie utilisée chez une femme enceinte, atteinte d'angiomatose de von Hippel-Lindau (AVHL) et de masses intracrâniennes.

Éléments cliniques : Une primipare de 30 ans, atteinte d'AVHL, qui désirait une analgésie épidurale pendant le travail, a été évaluée à la clinique d'anesthésie obstétricale à 35 sem de grossesse. Elle présentait des antécédents de céphalées et d'étourdissements. Un examen plus poussé a montré un hémangioblastome cérébelleux étendu et des effets de masse locaux significatifs. Un accouchement par césarienne combiné à une craniotomie de la fosse postérieure ont été réalisés à 37 sem de grossesse. Une anesthésie générale avec du fentanyl, du rocuronium, un mélange de protoxyde d'azote et d'oxygène ainsi que de l'isoflurane a été administrée pour la césarienne. Après l'accouchement, on a réduit l'apport d'isoflurane et amorcé une perfusion de propofol à 4–8 $mg \cdot kg^{-1} \cdot h^{-1}$. L'opération et la récupération se sont déroulées sans incident.

Conclusion : Pendant la grossesse de patientes affectées par un AVHL, des lésions préexistantes peuvent s'aggraver ou d'autres lésions peuvent se développer. Certaines, asymptomatiques, peuvent augmenter les risques de complications anesthésiques. On doit procéder à une évaluation complète de ces patientes avant l'anesthésie.

MANY patients with von Hippel-Lindau disease (VHLD) have hemangioblastomas of the cerebellum and spinal cord. While many patients have completed pregnancies without any complication, some develop new symptoms or have exacerbations of preexisting lesions during pregnancy.[1] A 30-yr-old primigravida with VHLD at 35 weeks gestation was found to have an enlarged cerebellar hemangioblastoma with significant local mass effects. There were evidence of uncal and trans-tentorial herniation as well as displacement of the brain stem. Due to the life threatening neurological condition, a craniotomy was deemed necessary at 37 weeks gestation. After a full discussion by the perinatal and surgical teams and with the consent of the patient, a Cesarean section delivery and a craniotomy were performed. We report the assessment and anesthetic management of this patient with VHLD and an intracranial mass lesion for Cesarean section delivery and posterior fossa craniotomy.

Case report

A 30-yr-old primigravida (height 160 cm, weight 60 kg) was referred to the preanesthesia clinic at 35 weeks

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gestation for assessment. The patient had VHLD diagnosed six years before. She was blind in the right eye because of retinal detachment and multiple retinal hemangioblastomas. She had undergone numerous photocoagulation procedures to treat lesions in the left eye. Previously, she had undergone three craniotomies at three, five, and six years for excision of cystic cerebellar hemangioblastomas and to manage obstructive hydrocephalus symptoms. Prior to pregnancy, renal tumours and pheochromocytoma were excluded by abdominal ultrasound and 24 hr urine collection for metanephrine. Cervical spine and brain magnetic resonance imaging (MRI) scan before pregnancy showed 11 cerebellar, brain stem and cervical spine (C3) hemangioblastomas (Figure 1). The largest, about 2 cm in diameter, was in the inferior aspect of the left cerebellar hemisphere and had mixed cystic and solid composition. There was no significant mass effect at that time.

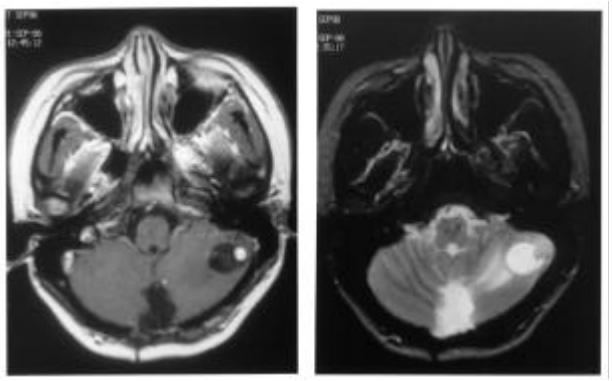

FIGURE 1 Brain-magnetic resonance imaging (MRI), three months before pregnancy. A) T2 not enhanced B) T1 postgadolinium enhanced, showing the largest hemangioblastoma with 1.8-cm cyst in the left cerebellar hemisphere.

At 30 weeks gestation, the patient began to experience more headaches and dizziness and a repeat MRI was planned after delivery. During the preanesthetic visit, the patient reported that she required six to eight tablets of acetaminophen (625 mg) per day for right-sided frontal headaches. The headaches were constant and mostly supra, and retro-orbital in distribution. The ophthalmologist suspected an intra-ocular tumour recurrence, recommended conservative management during pregnancy and to postpone further investigations until delivery had occurred. On examination, the airway, cardiovascular and respiratory systems were normal. There was no evidence of sensory deficit, motor weakness or gait disturbance.

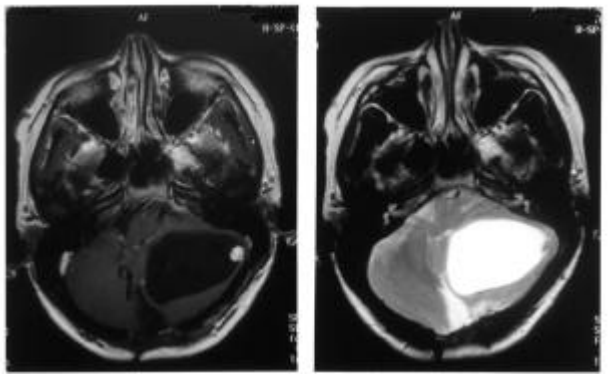

FIGURE 2 Brain-magnetic resonance imaging (MRI), at 36 weeks gestation: A) T2 weighted not enhanced, B) T1 weighted postgadolinium enhanced, showing the massive enlargement of the cystic (5.5 cm) component of the cerebellar tumour. There is left to right and posterior to anterior displacement of the brain stem.

At 36 weeks gestation, a repeat MRI scan of the brain and the spine showed multiple hemangioblastomas. The cystic component of the left cerebellar hemisphere lesion had increased in size to 5.5 cm in diameter (Figure.2). There was a significant local mass effect resulting in left-sided tonsillar herniation (Figure 3), mild upward trans-tentorial herniation of the superior cerebellar vermis, and displacement of the brain stem towards the right and anteriorly. The patient's clinical symptoms and radiographic findings indicated that the neurologic condition might be life threatening and urgent neurosurgical intervention was needed. The perinatal and neurosurgical teams discussed the options with the patient. A combined Cesarean section delivery and posterior fossa craniotomy at 37 weeks gestation was agreed on.

Preoperatively, the patient received ranitidine 150 mg, metoclopramide 10 mg and 30 ml 0.3 M sodium citrate solution. In the operating room, the patient was placed in a 15 head up position with a left lateral uterine displacement. Monitoring included pulse oximetry, noninvasive blood pressure measurement and a five lead electrocardiogram. Under local anesthesia and sedation with fentanyl 50 µg intravenously, a left radial arterial line, a left ante-cubital central venous pressure line, and a urinary catheter were inserted. Before induction of anesthesia, the patient's blood pressure (BP) was 155/85 mmHg, heart rate (HR) 92 beats per minute (bpm) and central venous pressure (CVP) was 11 mmHg. The patient received 100% oxygen by facemask and hyperventilated for

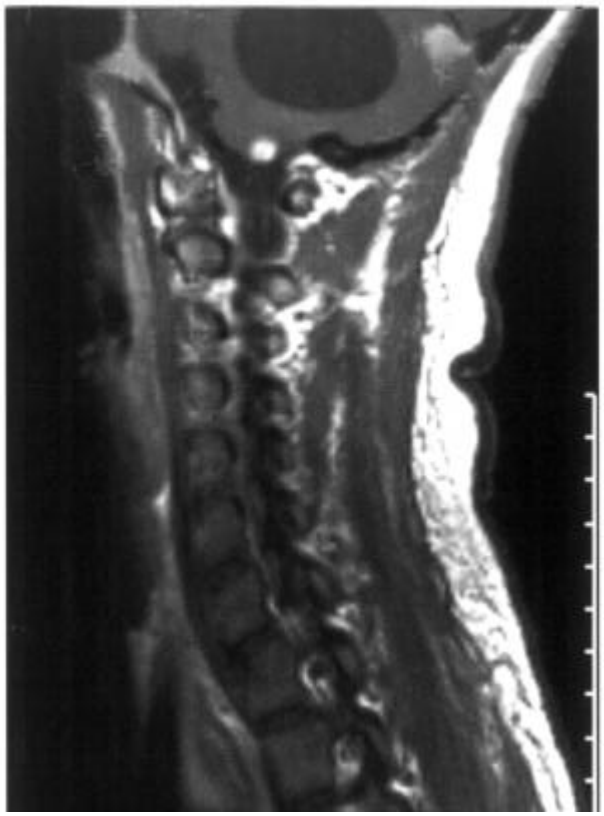

FIGURE 3 Sagittal T1 weighted postgadolinium enhanced magnetic resonance imaging (MRI), of upper C- Spine, and Foramen Magnum region at 36 weeks gestation, showing left-sided tonsillar herniation

three minutes. Anesthesia was induced with fentanyl 200 µg, sodium thiopental 300 mg, and rocuronium 50 mg. Cricoid pressure was applied upon loss of consciousness. The patient's lungs were manually ventilated to airway pressures of 15 cmH_2O with 100% oxygen and 2% isoflurane. Neuromuscular blockade was monitored with peripheral nerve stimulation of the right ulnar nerve. The trachea was intubated, anesthesia was maintained with N_2O: O_2 (50–50%) and 0.7% isoflurane (end tidal (ET)). Ventilation was adjusted to maintain $PaCO_2$ at 30 mmHg. During surgery, BP ranged between 130–110 / 80–65 mmHg, HR between 72–82 bpm and CVP between 8–10 mmHg. Four labetalol 10-mg boluses (total 40 mg) were used to maintain the blood pressure within the stated ranges.

A male baby with Apgar Scores of 5, 7, 9 at one, five, ten minutes was delivered three minutes after induction of anesthesia. The baby required one dose of *i m* naloxone (0.04 mg) for weak respiratory efforts two minutes after delivery.

After delivery of the infant, the patient received ampicillin 2 gm, midazolam 2 mg, and sufentanil 15 µg *iv*. An *iv* infusion of oxytocin 40 units·L^{-1} was initiated at a rate of 100 ml·hr^{-1}.

After closure of the uterus and abdomen, the patient was positioned in a three-quarter prone (park bench) position. Mannitol 40 gm and dexamethasone 10 mg were administered. Isoflurane was reduced to 0.4% (ET) and a propofol infusion was initiated at a rate of 4–8 mg·kg^{-1}·hr^{-1}, to minimize the risk of postpartum hemorrhage. Initially, the brain was tense and the neurosurgical dissection was difficult because of the three previous craniotomies. The largest cyst in the left cerebellar hemisphere was drained and resected. The estimated blood loss for the two procedures was 1200 ml, of which 400 ml were lost during the craniotomy. At the end of surgery, the patient was turned to the supine position. Neuromuscular block was reversed with neostigmine 3 mg, and glycopyrolate 0.6 mg. After transfer to the postanesthesia care unit, the propofol infusion was stopped and the patient received fentanyl 50 µg, as well as lidocaine 80 mg. Within ten minutes, the patient was awake and the endotracheal tube was removed without any coughing. The postoperative course was uneventful.

Discussion

VHLD

VHLD is a rare, autosomal dominant disorder that may be associated with retinal and cerebellar hemangioblastomas, renal cell carcinoma as well as pheochromocytoma. Patients with VHLD may also have cystic lesions of the epididymis, pancreas, kidney and other visceral organs. The causative gene has been localized to chromosome 3, and the use of linked markers has made prenatal diagnosis possible. The estimated incidence of VHLD is 1 in 35,000—65,000.[2] The clinical manifestations of the disease are variable. There are no symptoms in the fetal or neonatal periods. Ninety-five percent of patients develop clinical features before the age of 50, but the onset of symptoms in adolescence is rare.[3]

The retinal and central nervous system (CNS) hemangioblastomas are vascular units without endothelial tight junctions, so they leak plasma and blood. In 80% of the cerebellar lesions, the tumours are nodules in the wall of cysts. Some CNS lesions progress rapidly over a period of 2–4 yr, others progress slowly over many years, and some even become static.[3] The retinal tumours are associated with retinal exudate.[4] Even with early and aggressive treatment, as in our case, the retinal lesions can lead to glaucoma and blindness.

Most of the patients with VHLD have no problems during pregnancy but some may have worsening of existing lesions or develop new lesions. Grimbert *et al.*,[5] surveyed 30 women with VHLD, who completed 56 pregnancies. While most of the pregnancies had a favourable outcome, three of 56 pregnancies had complications. One patient developed intra-cranial hypertension at 32 weeks gestation from her cerebellar hemangioblastoma. The second patient developed eclampsia associated with pheochromocytoma at 32 weeks gestation. The third patient developed acute abdominal pain at 34 weeks gestation related to pancreatic cystadenoma. Joffe *et al*.[1] reported a 35-yr-old parturient with VHLD. She was diagnosed with pheochromocytoma at 22 weeks gestation, and required treatment with phenoxybenzamine and propranolol during her pregnancy. Ogasawara *et al*.[3] described a 23-yr-old patient who presented at 35 weeks of gestation with paraplegia secondary to acute intramedulary hemorrhage from a spinal hemangioma at the thoracic (T) 4–5 level. The patient had Cesarean section delivery under epidural anesthesia despite the presence of an asymptomatic lesion at T7–8 level. Further prospective studies are needed to determine how often complications develop during pregnancy in patients with VHLD.

Case presentation

The enlargement of the cerebellar lesion in our patient was an unexpected development. The extensive bilateral retinal lesions made it difficult to assess the presence of papilledema. The diagnosis was made with high index of suspicion and MRI.

Despite the patient's initial wish for a vaginal delivery, the medical staff were concerned about the stress of labour, fluctuations in blood pressure and further increases in intracranial pressure. With the symptoms of intracranial mass effect at 36 weeks, neurosurgical intervention was needed. One option was to perform the craniotomy, monitor the fetus in the perioperative period and deal with labour and / or complication if that occurred. We were concerned about the possible adverse effects of a prolonged anesthetic and surgery, as well as hemodynamic and metabolic changes on the fetus. If labour started during the craniotomy or within a short time after the craniotomy, the mother might suffer more complications. After full discussion by the perinatal and neurosurgical teams, and with the patient's consent, a combined Cesarean section delivery and posterior fossa craniotomy were performed.

Anesthetic management

The usual rapid sequence induction for general anesthesia in pregnant patient may not be tolerated in patients with raised intracranial pressure.[6] We used a non-depolarizing agent to achieve muscular relaxation in two to three minutes, while ventilating the lungs during that time. This allowed an appropriate depth of anesthesia with isoflurane, and maintained low arterial carbon dioxide level. The patient's favourable airway, absence of symptoms of gastro esophageal reflux, preoperative administration of ranitidine, non-particulate antacid and metoclopramide, and application of cricoid pressure decreased the risk of pulmonary aspiration. Bédard *et al*.[7] reported on anesthesia for Cesarean section for a parturient with an acoustic neuroma compressing the pons, midbrain and fourth ventricle. The authors applied cricoid pressure as consciousness was lost and ventilated the patient's lungs with positive pressure by mask before tracheal intubation. There is little evidence in the literature to indicate whether any approach is preferable.

In summary, patients with VHLD may have worsening of preexisting lesions or develop other lesions during pregnancy. Some asymptomatic lesions can increase the risk of anesthesia complications. These patients need a comprehensive assessment before the administration of anesthesia.

Acknowledgement

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