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Three anesthetic challenges for spinal surgery in a morbidly obese achondroplastic dwarf.

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Summary : We report the anesthetic management of a patient who was achondroplastic, morbidly obese (BMI 66.7 kg/m²) and required surgery in the prone position. Thanks to effective teamwork and preparation, these three challenges (short stature, morbid obesity and prone position) were overcome using the Cotrel operating table, ultrasonography and flexible bronchoscopy.

Keywords : spine surgery ; achondroplastic dwarf ; Cotrel operating table.

Introduction

Achondroplasia is the most common form of short limbs dwarfism, affecting about one in 20,000 newborns. It is caused by a mutation of the fibroblast growth factor receptor-3 gene. This mutation interferes with bone formation and has a detrimental effect on craniofacial and vertebral development. It can as well induce hydrocephalus, compression of the spinal cord or nerve roots. Most cases of achondroplasia occur as a result of de novo mutations, but some may be transmitted in an autosomal dominant pattern of inheritance (1, 2, 3).

We present a case of a morbidly obese achondroplastic patient suffering from sleep apneas. The patient was scheduled for a lumbar laminectomy L1-L5 in the prone position.

For publication of this report, the patient's written consent was obtained.

Description

This 59-years old achondroplastic woman was admitted for multi-stage (L1-L5) spinal stenosis surgery. She had been morbidly obese for decades, and had gained more weight after developing an invalidating spinal stenosis. Her body mass index was 66.7 kg/m² (96 kg for 120 cm). She had severe sleep apnea syndrome (SAS) treated by CPAP and oxygen (2L/min) during nighttime to maintain SpO_2 between 80 and 90%. She also had restrictive pulmonary disease and orthopnea. She had already undergone tonsillectomy and hysterectomy (2002) at another institution. The family reported difficult airway management but the anesthetic records were not available.

Physical examination revealed an obese achondroplastic woman with short limbs and a large skull with frontal protrusion (Fig. 1). Hemoglobin saturation when breathing room air was 94%. Her clavicles were deeply situated in a high position in a short neck partially covered with redundant adipose tissue. Therefore, central venous access in this part of the body was considered difficult and dangerous to obtain.

Airway examination yielded normal neck extension and mouth opening, a Mallampati score 3, and a chin-thyroid distance of 5 cm. The patient could remain supine during the examination. She had a plethoric but depressible abdomen. Peripheral venous access was very poor but we were able to insert a 20 G IV cannula in her hand to obtain preoperative blood samples.

Routine laboratory results and electrocardiogram were without any particularities.

Because of the risk of pulmonary arterial hypertension secondary to SAS, transthoracic echocardiography was performed. However, obesity and shape of thorax did not allow the evaluation of the pulmonary pressures. Therefore, we decided to

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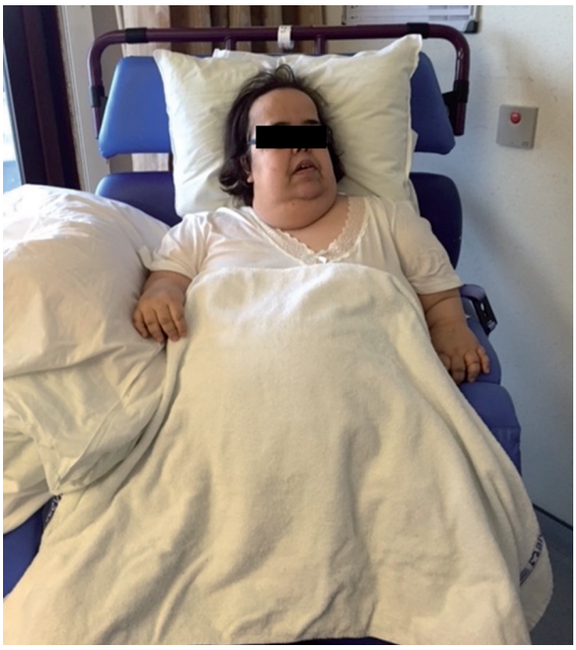

Fig. 1. — Achondroplastic Dwarf.

consider the patient as if at high risk for pulmonary arterial hypertension and adapted anesthesia management accordingly avoiding hypoxemia, hypercapnia and acidosis. Inhaled Nitric Oxide was also available in the OR if needed.

In the operating room, we installed the patient on a large table, with a mild anti-Trendelenburg position. The patient was monitored with pulse oximetry and a 5-lead ECG. Effective preoxygenation was achieved by facial mask. We inserted a catheter for invasive blood pressure monitoring before inducing anesthesia because non-invasive blood pressure measurements were inconsistent due to shortness of the limbs. Several attempts to cannulate the radial arteries were performed under ultrasound guidance but the catheters kinked in the subcutaneous tissue and failed to show an arterial waveform only a few minutes after their placement. Finally, we used the femoral route to introduce both a central venous (Arrow Blue Line® 7F 20 cm, double lumen catheter) and arterial (Seldicath® 5F) access under ultrasound guidance.

General anesthesia was induced with sufentanil (5 μg) and propofol (150 mg). We chose to use a small opioid dose to prevent the risk of postoperative apnea in this susceptible patient. After loss of consciousness, four-hand ventilation with a classic facial mask was performed easily. A dose of rocuronium 0.9 mg/kg (ideal body weight) was administered to obtain good intubation conditions rapidly. Laryngoscopy with a MacIntosh blade 3 and without any hyperextension of the neck

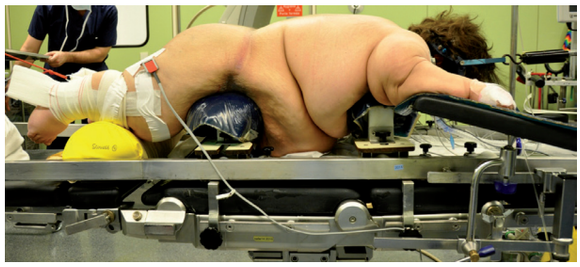

Fig. 2. — Prone position on the Cotrel operating table.

revealed a Cormack-Lehane grade I glottic vision and a 7.0 mm ID cuffed endotracheal tube (Sims-Portex®) was inserted. Correct positioning of the tube was verified using a fiberoptic bronchoscope to avoid any selective ventilation due to the shortness of the tracheobronchial tree.

Then, the patient was installed in prone position for surgery. A Cotrel table was used in order to limit compression of the thorax and the abdomen which could jeopardize venous return. A cranial halo was inserted in the skull without traction and two adhesive tapes were placed on the lower limbs (Fig. 2).

Maintenance of anesthesia consisted of sevoflurane (Sevo Et 2-2,2%), iterative boluses of sufentanil 5 μg (total dose: 15 μg) and administration of iterative doses of rocuronium (total dose: 100 mg). A NeuroSENSE® (NeuroWave System Inc) was used to monitor depth of anesthesia.

The intraoperative period was uneventful. The patient was extubated immediately, after checking that reversal of muscle relaxation was adequate (TOF ratio > 0.9).

Immediate postoperative analgesia consisted in IV paracetamol 1gr, IV ketorolac 30mg and the titration of 2 mg/2 mg of piritramide (Dipidolor®) (total dose: 8 mg), a synthetic opioid equipotent to morphine. At the end of the procedure, the patient was able to move her legs on demand, was transferred to the ICU and stayed there 72 hours, essentially because of respiratory acidosis (worst pH and $paCO_2$ were respectively 7.23 and 61 mmHg) due to severe postoperative apneas. These apneas were treated with several sessions of bilevel positive airway pressure during daytime and continuous positive airway pressure with oxygen supply during the night.

Discussion

This patient presented with severe movement restriction due to lumbar stenosis. Endochondral bone formation is affected by the disease and

results in abnormally shallow vertebral bodies and underdeveloped vertebral arches. The spinal canal is thus constricted throughout its length and leads to narrowing of the subarachnoid and epidural spaces. Moreover, intervertebral discs protrusion is common in dwarfs. The association of spinal canal stenosis and disc prolapse explains the frequent symptoms of neural compression in these patients (4).

Obesity is current in achondroplastic dwarfs, with an incidence 2 to 8 times higher than in the general population. Moreover, dwarfism and obesity share several synergistic comorbidities, especially on upper airways, sleep-disordered breathing and pulmonary function: they all need to be evaluated and anticipated for safe anesthetic management (1). Moreover, hyperextension of the neck should be avoided in achondroplastic patients, due to the possibility of cervical cord compression. To prevent ventilation and intubation difficulties during the induction of anesthesia, thorough clinical examination of mandibular mobility, size of the tongue, mouth and mandible, shortness and mobility of the neck need to be evaluated. The results of clinical assessment should be confirmed by a radiological evaluation of the cervical spine and the skull (radiographs, CT-scan or MRI techniques) (1, 4). Furthermore, all available previous anesthetic records should be reviewed for information about airway management, such as difficulty with facial mask ventilation or with classic laryngoscopy (even with a short handle) and size of the endotracheal tube. However, in a series of 36 anesthetic procedures on achondroplastic patients, Mayhew *et al*. (5) did not encounter ventilation or intubation difficulties under general anesthesia. Furthermore, in our case, we did not have to deal with difficult airway management. Besides, due to the shortness of the tracheobronchial tree, the size and length of the endotracheal tube is smaller than would be expected by the age of the patient. We thus verified the correct position of our endotracheal tube with a flexible bronchoscope (1, 6).

Sleep-disordered breathing is the most frequent comorbidity in achondroplasia (7). On one hand, upper airway obstruction is related to abnormal craniofacial bone growth, with brachycephaly, pharyngeal hypoplasia, flattening of the nasal bridge and to hypotonia of the upper airway muscles. On the other hand, central sleep apnea could be caused by compression of the distal medulla and the cervical cord by stenosis of the foramen magnum (4). Recent guidelines recommend a systematic polysomnography for these patients (8). Even more so, Julliand *et al*. (2) confirm a high prevalence of polysomnographic abnormalities (93%) and a poor correlation between the patient's symptoms and the objective abnormalities on polysomnography. Our patient presented both obstructive and central apneas and needed a CPAP device to sleep. Her prolonged stay in the ICU after surgery was mainly due to severe apneas with respiratory acidosis.

Because of progressive thoracic kyphosis, some patients develop a restrictive pulmonary dysfunction with reduction in vital capacity, a low functional residual capacity and a high closing volume. In these cases, the results of the preoperative spirometry should be interpreted with caution because there are no standard references in the literature in short-trunk dwarfs. However, although the lungs may be smaller, they are usually functionally normal. Furthermore, thoracic dysplasia seems to be a problem essentially confined to young children. Indeed, Stokes *et al*. (9) demonstrated no repercussion in achondroplastic adults, except for a non clinically significant reduction of the anteroposterior chest diameter in men.

Chronic upper airway obstruction, sleep-disordered breathing and restrictive pulmonary dysfunction all contribute to the development of pulmonary hypertension and eventually chronic cor pulmonale. These are the most common cardiovascular disturbances in achondroplasia. They should be evaluated preoperatively. Unfortunately, in our case, we assumed that she had some pulmonary hypertension because the cardiologist was unable to evaluate the pulmonary pressures by transthoracic echocardiography and because we could not use transesophageal echocardiography during the surgery due to the ventral positioning. Preventing acute elevation of pulmonary arterial pressures during the procedure is important because this could lead to right-sided heart failure, decreased cardiac output, myocardial ischemia and acidosis. (4)

Another particularity of achondroplasia is frequent unsuccessful venous access with an incidence between 10 and 50%, due to excess of subcutaneous tissue, flaccid skin and joint deformities (1, 6). For this reason, we used ultrasound guidance for vascular access.

In conclusion, with effective teamwork and preparation, the three challenges (short stature, morbid obesity and prone position) have been overcome using the following three tips: Cotrel table, ultrasonography and fiberoptic bronchoscopy.

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