ANESTHETIC MANAGEMENT OF ACHONDROPLASTIC DWARF UNDERGOING CESAREAN SECTION

- A Case Report -

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Abstract

There are more than 100 different types of dwarfism. Achondroplasia is the most common of these conditions. The aim of this report is to describe the anesthetic management of these patient, discussing the anesthetic considerations and emphasizing the difficulties encountered. A 32-year-old achondroplastic parturient underwent cesarean section under general anesthesia. We did not encounter problems related with airway management. The operation went without any complication. There are risks for both regional and general anesthesia in achondroplastic patients. The most important point is the careful preoperative assessment. Anesthesia plan should be specified to individual basis.

Introduction

Various uncommon disorders present in pregnant women can create a dilemma for obstetricians and obstetric anesthesists as to how to best manage these patients. Achondroplastic dwarfism is one of these disorders and the clinical management remains controversial[1]. Achondroplasia is the commonest form of dwarfism, in which a large number of cases resulted from spontaneous mutation. The incidence varies between 4 and 15 per 100,000 live births. Females are affected more frequently than males[2].

Achondroplastic patients present several problems for both general and regional anesthesia. We describe a patient with achondroplasia undergoing Cesarean section and discuss the anesthetic considerations in achondroplastic individuals.

Case Report

A 32-yr-old primagravid achondroplastic dwarf presented for elective Cesarean section at 37 weeks' gestation. She had no previous history of anesthesia and her pregnancy had been uneventful. Her history included no relevant conditions. She was taking prenatal vitamins and had no known allergies.

Physical examination revealed a 49-kg, 135 cm, normal intelligent female with large head, short limbs and mild kyphoscoliosis. She had short neck, large tongue, protruded chin, full set of teeth. Mouth opening was adequate with Mallampati II airway and neck extention was not limited. Examination of cardiorespiratory system revealed no problems and there was no neurological abnormalities. Preoperative hemoglobin and electrolyte concentrations were within normal limits. Ultrasonic measurements in utero suggested that the fetus was normally developed.

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The patient was very anxious and she did not desire to be awake during procedure. Therefore, after discussion of potential risk and written informed consent, it was decided to performe the operation under genral anesthesia. Appropriate sized facemasks, tracheal tubes, stillettes, laryngeal mask airways (LMA™) and intubating laryngeal mask airways (ILMA™) were prepared for possible difficulty in intubation.

No premedication was given and she was monitored by non-invasive arterial pressure, electrocardiogram (ECG) and pulse oxymetry on arrival in the operating room. Anesthesia was induced with thiopenthal and neuromuscular blockade was achieved with succinylcholine. Trachea was intubated at the first attempt with a 7.0-mm ID endotracheal tube. Lower segment cesarean section proceeded and a live baby was delivered weighing 3200 grams with APGAR scores of 8 and 9 at the first and the fifth minutes respectively. Maintenance of anesthesia was provided by sevoflurane in 50% oxygen-nitrous oxide mixture. Muscle relaxation was achieved with vecuronium the patient was given oxytocin and fentanyl after the umbilical cord was clamped. No other medication was given during the procedure. The total duration of anesthesia was 1h and all monitored parameters (ECG, pulse oxymetry, endtidal CO_2 and non-invasive blood pressure) remained stable. At the end of the operation, neuromuscular blockade was reversed with neostigmine 0.05 $mg.kg^{-1}$ and atropine 0.02 $mg.kg^{-1}$ and the trachea was extubated when the patient was fuly awake. The patient was taken to postanesthesia care unit and administered 5 $l.min^{-1}$ of oxygen through a face mask until transferring to the ward. The postoperative period was uneventful and the patient and neonate were discharged on the fifth day.

Discussion

Achondroplastic dwarfs characteristically have low fertility rates; however, they often require delivery by cesarean section because the normal-sized fetal head and smaller than normal maternal pelvic diameter result in cephalopelvic disproportion during the later stages of pregnancy[3]. These patients have a number of anatomic and physiological abnormalities that contribute to problems with the administration of obstetric anesthesia.

Achondroplastic patients have facial features that alert the anesthesists to potential problems in airway management[4]. They have narrow nasal passages and pharyngeal and maxillary hypoplasia. Maintenance of a patent upper airway may be difficult because of the presence of a large tongue and mandible. The base of the skull is shorted (because of early fusion of constituent bones) and angulated, yielding a limited extension and making endotracheal intubation potentially difficult[1,5]. On the other hand, not all authors have encountered difficulties. Mayhew et al.[6] reported no difficulty in airway management or direct laryngoscopy in a series of 27 patients undergoing 36 anesthetic procedures. They also suggested that is tracheal intubation was required, a small tracheal tubes should have been selected. There are reports describing the achondrplastic patients with classical symptoms and signs of upper airway obstruction but no difficulty encountered[7-11].

We elected to use general anesthesia for two reasons. First of all, the patient wanted to be asleep because of her anxiety. Secondly, preoperative examination suggested no possibility of a difficult airway. Consequently, the intubation was completed in the first attempt without any undesirable effects.

Several anatomical abnormalities found in achondroplasia may complicate regional techniques. Difficulties may result from kyphoscoliosis and other spinal abnormalities, included narrowing of the vertebral canal, shortening of the pedicles, reduced interpedicular distance (particularly in the lower lumbar spine) and osteophyte formation. Spinal stenosis may impair cerebrospinal fluid flow, such that identification of dural puncture is more difficult. Also the presence of narrow epidural space make insertion of catheter difficult[12]. Engorged epidural veins increase the risk of venous puncture either by the Tuohy needle or the catheter, and result in an unpredictable spread of local anesthetic within the space[13].

We found reports related with achondroplasia which had successfully managed by regional anesthesia. These are spinal[14-17], epidural[3,5,13,18-20] or combined spino-epidural (CSE) techniques[4].

In these reports, technically challenging problems owing to skeletal abnormalities and the appropriate

dose of drugs were discussed. Ravenscroft et al.[16] were successful in one patient with the spinal anesthetic dose decreased by 30%. DeRenzo et al.[17] also reduced the intracheal dose of bupivacaine from 12 to 10 mg with the addition of 0.2 mg morphine for the relief of post-cesarean section pain[21]. In that report, the patient had experienced discomfort, requiring iv sedation for forty minutes after delivery. The suggested that decreasing the intracheal dose was not unreliable.

It's suggested that epidural anesthesia was theoretically preferable to spinal anesthesia because of ability to titrate the level of block. Incremental doses of anesthesia could be required due to maternal short stature and kyphoscoliosis[1,4]. Trikha et al.[4] preffered CSE over a spinal or epidural technique because it combines the rapid onset of spinal anesthesia with the implantation of epidural catheter. This approach provided prolonged anesthesia and excellent postoperative analgesia. DeRenzo et al.[17] also reported that titratable techniques such as epidural, CSE, continuous spinal would have been beter suited for these paitents.

In conclusion, the risks of both regional and general anesthesia in achondroplastic patients are known. With the above factors taken into consideration, a complete history and physical examination before administration of anesthesia can help to reduce risks. The anesthesia plan should be based on each indivdual case and the potential risks must be discussed with the patient.

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