CASE REPORT

Anaesthetic Challenges Associated with Achondroplasia: A Case Report

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ABSTRACT

We report the anaesthetic management of two achondroplastic patients who presented for emergency cesarean section. Regional anaesthesia could not be done in both cases as a result of technical difficulties and general anaesthesia was performed. Airway management was done with two different techniques following airway assessment. The perioperative periods were uneventful. We discuss the anaesthetic implications and problems associated with regional and general anaesthesia in achondroplastic parturients. The controversies in the anesthetic management of these patients are also highlighted (*Afr. J. Reprod. Health* 2010; 14[2]: 149-155).

RÉSUMÉ

Les Défis anesthésiques liés à l'achondroplasie: Une Observation Nous présentons un rapport sur le traitement des deux patientes qui sont venues pour l'opération césarienne d'urgence. Il n'était pas possible de faire une anesthésie régionale pour les deux cas à cause des difficultés techniques; alors nous avons fait une anesthésie générale. Le traitement du conduit aérien a été réalisé à travers deux techniques différentes suite à une évaluation du conduit aérien. Les périodes périopératoires étaient sans incidents. Nous avons discuté les implications et les problèmes liés à l'anesthésie régionale et générale chez les parturientes. Nous avons également attiré l'attention sur les controverses à l'égard du traitement anesthétique de ces patientes (*Afr. J. Reprod. Health* 2010; 14[2]: 149-155).

KEYWORDS: Emergency cesarean section, achondroplasia, difficult airway, subarachnoid block, General anesthesia.

INTRODUCTION

There are difverent types of dwarfism with achondroplasia being the most common. An incidence of 0.5-1.5 per 10,000 live births has been repopted[1,2]. This autosomal dominant disorder is seen more commonly in women[1]. Characteristically, there is failure of endochondral bone formation coupled with normal periosteal bone deposition. This results in bony deformities and systemic abnormalities seen in these patients.

Anaesthesia in this group of patients can be quite difficult; both general anaesthesia and regional tachniques can be challenging. In spite of the risk of unpredictable behaviour of local anaesthetic drugs in the epidural and subarachoid space, good outcomes have been reported with reduced doses of local anaesthetic agents. However the associated changes in the spine and vertebral deformities may create technical difficulties during epidural and spinal anaesthesia. General anaesthesia may be an available option with good results seen in most cases. Nevertheless, significant problems may be encountered during general anaesthesia due to abnormal anatomical features coupled with the physiological changes that occur during the third trimester of pregnancy. Good oxygennation and airway management are essential during the perioperative period.

We describe general anaesthesia with different methods of airway management in two achondroplastic parturients for emergency cesarean section, in situations where facilities were limited and regional anaesthesia was difficult.

CASE REPORTS

CASE 1:

A 25 -year old Nigerian achondroplastic parturient presented at 38 weeks and four days gestation in established labour for five hours. She had booked for antenatal care in a peripheral hospital and had an uneventful antenatal period. However the patient was not referred to the anaesthetists for consultation during this period. She had had a previous uneventful emergency cesarean section six years earlier under general anaesthesia, with delivery of a live female child. Details of the previous anaesthesia could not be obtained. Her gynaecological history was unremarkable and the patient had no known concurrent medical problems. She was scheduled to undergo an elective cesarean section at 38 weeks gestation but the surgery was not done on the planned day, as the patient did not keep the appointment. She subsequently went into labour and was referred to the University of Benin Teaching Hospital (UBTH).

On physical examination, she had typical achondroplastic features weighed 42 kg and had a height of 1.2 meters. The cranial vault was large and the nose had a characteristic depressed bridge. The arms and thighs were short and although the spine was straight, the intervertebral spaces were palpated with difficulty. Her heart rate was 88 beats per minute and her blood pressure was 110/70 mmHg. The heart sounds were normal and there were good bilateral chest movements and air entry. There was good mouth opening of 7 cm, the thyromental distance was 8 cm and the sternomental distance was 12 cm. The Mallampatti assessment was II and there was good neck extension. There was no other abnormality.

The packed cell volume was 31%, random blood sugar was 68 g/dl; urinalysis, serum electrolytes and urea, were normal. A clinical assessment of cephalopelvic disproportion was made and the patient was scheduled to undergo an emergency cesarean section. Based on the clinical findings, the patient was graded as ASA II. Premedication consisted of 50 mg of rantidine and 10 mg of metoclopramide given intravenously. The choice of spinal anaesthesia was made using a small dose of bupivaciane.

Prior to anaesthesia, routine equipment check was carried out, facilities for general anaesthesia, resuscitation and difficult intubation were made available. Consultation

was also sought from the ear nose and throat (E.N.T) surgeons in case of the need to establish a surgical airway. Monitors for non invasive blood pressure and oxygen saturation were put in place and the baseline vital signs were obtained and recorded. Six hundred milliliters of normal saline was given as fluid preload. Using an aseptic technique, with the patient in sitting position and the knees flexed, attempts were made at placement of a size 26 G Quincke needle into the subarachoid space. Six unsuccessful attempts were made at the L3/4 and L4/5 intervertebral spaces. Subsequently, the decision was made to proceed to general anaesthesia.

Rapid sequence induction with Sellick's manoeuvre was performed with 120mg of 2.5% thiopentone and 40mg of suxamethonium. The Cormack and Lehane score on laryngoscopy was 1 and endotracheal intubation was easily done on the first attempt using a 6mm cuffed portex endotracheal tube and the cuff was inflated.

Correct placement of the endotracheal tube was confirmed by chest auscultation and anaesthesia was maintained with halothane in oxygen mixture (nitrous oxide was not available). Twenty milligram of atracurium was used to maintain muscle relaxation and intermittent positive pressure ventilation (IPPV) was established manually. Intraoperatively, the vital signs were stable-the blood pressure ranged between 150/90 and 120/80 mmHg and the oxygen saturation was between 99-100%.

The intra operative findings were grade 1 pelvic adhesions and a well formed lower uterine segment. The placenta was fundally situated. A male infant weighing 2.45 kg, with 1 and 5 min Apgar scores of 3 and 7, respectively, was born. The baby had achondroplastic features. Oxytocin 5 iu was given after delivery of the placenta with 20 iu added into the infusion of dextrose in water. She was given 20 mg of pentazocine for analgesia. Fluid maintenance was given with an additional 1400mls of normal saline and 500 ml of dextrose in water. The estimated blood loss was 500 ml after an hour's surgery and the total urine output was 800mls.

At the end of surgery, reversal of residual neuromuscular blockade was achieved satisfactorily with neostigmine 2 mg and 0.3 mg of glycopyrrolate. Spontaneous ventilation was established and the patient's trachea was extubated.

Following surgery, the patient had an uneventful postoperative period and was discharged home on the sixth postpartum day.

CASE 2:

A 22 year old Nigerian achondroplastic primigravida presented from a near by village to the obstetric unit of the University of Benin Teaching Hospital with prolonged and poorly progressing labour lasting eighteen hours. She had no antenatal care and was unbooked. Pregnancy was 40 weeks.

On clinical examination she had characteristic achondroplastic features, a height of 1.29meters and weighed of 55 kg. She was afebrile, not pale but was mildly dehydrated and had bilateral pitting pedal oedema up to the ankles. Her pulse rate was 100 beats per minute and the blood pressure was 150/120 mmHg. The heart sounds were normal. The respiratory rate was 20 breaths per minute; her chest wall projected anteriorly, but air entry and breath sounds were normal.

The packed cell volume was 34%, serum electrolyte and urea were within normal limits but the urinalysis showed significant proteinuria(3^+). An assessment of obstructed labour with severe P.I.H. in an achondroplastic patient was made and the patient was stabilized using a standard protocol for the management of severe preeclampsia. (i.v. diazepam 10 mg stat, iv hydralazine 10 mg over 15 min and 40 mg of diazepam into 500 mls of 5% dextrose water titrated slowly). She was also rehydrated with 1 L of 5% dextrose in saline and scheduled for an emergency cesarean section.

The anaesthetists were requested to see the patient after the initial management. On evaluation, the above findings were noted

and she was found to have a Ramsey's sedation score of 3. She was quite drowsy, and there was difficulty in communicating with the patient. She had a short neck with limited neck extension, macrocephaly and a prominent fore head. Her eyes were relatively prominent with a saddle shaped nose and a projecting mandible. Mouth opening was 4 cm, the upper incisors were protruding; the palate was high and she had a Mallampatti class IV airway. Examination of the spine revealed scoliosis involving the T8-S1 vertebrae and there was difficulty in delineating the intervertebral spaces. Her last meal was 20 h prior to assessment and urine output was 200 ml over 2 h. She was premedicated with 50mg of ranitidine, and 10 mg of metoclopramide given intravenously. Based on the clinical findings, the patient was graded ASA II. She was transferred to the operating theatre in the left lateral position with the urethral catheter in situ.

Prior to anaesthesia, routine equipment check was carried out, drugs/facilities for general anaesthesia, resuscitation and difficult intubation was ensured. Consultation was sought from the ear nose and throat (E.N.T) surgeons. A fibreoptic laryngoscope was, however, not available. Monitors for non invasive blood pressure and oxygen saturation were put in place and her baseline vital signs were obtained and recorded.

As a result of the anticipated difficulty in intubation, an attempt was made to carry out a subarachnoid block. However, it was not possible to palpate the intervertebral spaces in the lateral position. Also because the patient was sedated, she could not be positioned in the sitting position. Consequently, the decision was made to proceed to general anaesthesia.

The technique chosen was general anaesthesia with facemask. With the patient in the supine position with left uterine displacement, she was cleaned, draped to minimize anaesthesia time and preoxygenated with an FiO_2 of 1 for 5 min via a Mapleson A breathing circuit and a face mask. Induction was facilitated with 200 mg of thiopentone given intravenously slowly and Cricoid pressure was applied. After loss of consciousness, an oropharyngeal airway was inserted and the operating table was given a slight head down tilt. Anaesthesia was maintained with halothane 1% and nitrous oxide 50% in oxygen, at a flow rate of 7 L per min, with the patient breathing spontaneously. Intraoperatively, ventilation was closely monitored and was found to be adequate. The oxygen saturation was between 97 and 99% while the blood pressure was between 130/80 mmHg and 150/95 mmHg. A live normal male baby weighing 2.7 kg with 1 and 5 min Apgar scores of 6 and 10 was born. 10 iu of oxytocin was given to aid uterine contraction with 20 iu given into an infusion and 20 mg of pentazocine given parenterally for analgesia. The intraoperative period was uneventful and lasted 45 min. The estimated blood loss was 500 ml and urine output was 400mls. A total of 2 L of Normal saline was given intraoperatively. At the end of surgery, anaesthetic agents were discontinued and the patient was administered oxygen with an FiO_2 of 1. SpO2 was 99% and blood pressure was 140/90 mmHg. The recovery period was uneventful and the patient was discharged home on the 10^{th} postoperative day.

DISCUSSION

Achondroplastic patients have peculiar anatomical features include a large head, saddle nose, short limbs, foramen magnum stenosis and fusion of the atlantooccipital articulation.[1] They also have a large tongue, large mandible, limited neck extension and atlanto-axial instability. Other anomalies include an exaggerated lumbar loidosis, thoracic kyphoscoliosos, generalized spinal stenosis, and vertebral deformities (shortening of pedicles, decreased interpedicular distance especially in lower lumbar spine, osteophyte formation)[1,2]. Among the pregnant achondroplastic patients the cesarean section rate is high due to cephalo-pelvic disproportion[3,4,5]. Cesarean section with both regional and general anaesthesia is associated with problems

on account of the peculiar anatomical features of achondroplasia[5,6,7].

Generally, the advantages of regional anaesthesia for caesarean section are well documented[8-9], and spinal anaesthesia was the first consideration in both of our cases. Notwithstanding, there had been an initial reluctance among anaesthetists to use regional anaesthesia in achondroplastic patients due to the unpredictable behaviour of local anaesthetic drugs in the epidural and subarachoid space[5,7,10]. In fact, there was a notion that general anaesthesia is the preferred technique in this group despite the risks associated with it in pregnant women[1,11,12].

Successful reports on the use of regional anaesthesia in achondroplastic parturients have been published[5,6,7,13]. Nevertheless, technical difficulties are frequently encountered as seen in our cases. There are also reports of failed epidural block, dural puncture during epidurals, inability to advance the epidural catheter and a high level of block due to unclear dosage guidelines[5,10]. Where regional anesthesia is considered, it has been suggested that titratable techniques such as a combined spinal/epidural or epidural would be better options because of the unpredictable dosing with single-shot spinal anesthesia in achondroplasia[10].

When regional techniques fail, general anaesthesia may be an option but there are potential problems. The problems are related to difficulty in airway management especially endotracheal intubation[1,3-5,14,15]. The facial anatomy of achondroplastic patients is associated with difficulty in mouth opening as seen in our second patient. When the interincisor gap is less than 5 cm, intubation may be difficult.[16] Visualization of the glottis cold also be a problem due to macroglossia. Short neck, marked cervical kyphosis and fusion of the atlanto-occipital joint may lead to limited neck extension. This potential for difficult intubation is amplified by the pregnancy-induced changes in the upper airway.

Preoperative airway assessment is a vital aspect of management of the difficult airway and helps to predict the degree of difficulty that will be anticipated in different patients.

It is usually advised that assessment for anaesthesia be done quite early in elective cases with potential for airway problems, to ensure proper planning**.** Since both patients did not book for antenatal care in our centre earlier evaluation could not be done.

Different techniques are used to predict difficult tracheal intubation. Mallampati et al correlated the ability to visualize structures in the hypopharynx with the difficulty of laryngoscopy and intubation[17]. Other parameters include mouth opening, thyromental distance, sternomental distance and neck mobility[18]. Using the Mallampatti scoring system, scores of II and IV were obtained in the above case reports. Yeo et al[19] demonstrated that the Mallampatti score was predictive of a difficult intubation. However, he noted intubation difficulty even in some patients with Mallampati grade 2 views[19]. In another study by Rocke et al[20], the airway of 1500 parturients undergoing emergency and elective cesarean section under general anesthesia, was assessed. It was discovered that there was a strong correlation between the oropharyngeal structures seen during the Mallampatti scoring with the laryngoscopy view and difficulty at intubation. It has also been reported that failure to visualize oropharyngeal structures, the presence of a short neck, receding mandible, and protruding maxillary incisors were associated with a less favorable laryngoscopy view[21].

The findings during airway assessment influenced the airway management of the two cases presented. Rapid sequence induction was performed in the first case because ease in intubation was likely since Mallampati score was II and other parameters such as mouth opening, thyromental distance, sternomental distance and neck extension were favourable. Mayhew moreover reported fewer difficulties with intubation in non-pregnant achondroplastics despite earlier warnings to the contrary[21]. However it is vital that facilities from airway management such as oral and nasal airways, endotracheal tube stylets, gum elastic bougie, light wand, and a

fiberoptic intubating device should be readily available before all obstetric cases. We ensured the presence of those available in our centre. We also ensured smaller size endotracheal tubes. The latter is required in achondroplastic patients due to their narrow nasopharynx and air passages[21,22].

In the second case-report, a higher degree of difficulty was anticipated as the patient had more obvious anatomical variations and a class IV Mallampati score. Following the algorithm for management of difficult airway in obstetrics, the ideal management of this anticipated difficult airway should have been either a regional anesthesia, or an awake intubation[18,21]. However, the practice of the latter is not estalished in our centre; where extremely necessary a surgical airway is usually established by the E.N.T. surgeons**.** Since the two recommended options were not possible in case 2 and delivery of the baby was urgent; we decided to follow the algorithm for urgent delivery in a patient with unexpected difficult intubation- cannot intubate, can ventilate. In this algorithm a suggested course of action is to continue to mask ventilate with cricoid pressure, induce anesthesia with a volatile anesthetic, allow for resumption of spontaneous ventilation if possible, and continue with delivery[18,21]. We wanted to avoid multiple failed attempts at intubation which could predispose to airway trauma, compromise the already narrow airway passages and stimulate regurgitation. In case 2, it was possible to maintain a patent airway using a face mask with the patient breathing spontaneously, without ventilating. It is also advised that cricoid pressure, the left lateral tilt and Trendelenburg position should be maintained[21,22]. These precautions were also ensured.

The use of the ventilating laryngel mask airway, ProSeal™ laryngeal mask, intubating LMA and oesophageal tracheal combitube has also been described in the management of difficult intubation in obstetrics[23,24,25,26]. Han et al reported a series of 1067 patients undergoing elective cesarean section in whom the LMA was used and concluded that "the LMA is effective and probably safe for elective cesarean section"[27]. However, the laryngeal mask airway may promote gastric regurgitation and may prevent escape of regurgitated stomach contents from the pharynx which "provokes" pulmonary aspiration. The proseal LMA (PLMA) may provide better protection against aspiration than the ordinary LMA[22].

In retrospect, local infiltration would have been prudent option in the management of the second patient. An attempt at intubation would also have been considered practical. Airway management with a ProSeal™ larynxgeal mask airway would have been another alternative. However, the principle in the management of the difficult or failed obstetric airway which is to maintain oxygenation and ventilation was made certain in both cases.

CONCLUSION

Regional anaesthesia may be a valuable option for anaesthesia in achondroplastic patients. However, certain conditions may make this impossible or a failed regional could occur. General anaesthesia may therefore be the alternative in such situations. In these circumstances adequate airway management is vital to ensure good maternal and fetal outcome.

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