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Case Report

Anesthesia management of patient with achondroplasia for abdominal hysterectomy

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ABSTRACT

Achondroplasia is the commonest form of dwarfism. These patients present several problems for both general and regional anesthesia. There are reports describing regional anesthesia in such patients but there are scarce reports of general anesthesia. The aim of this report is to discuss anesthetic considerations in such patients and to emphasize the difficulties encountered. A 34 year old patient with achondroplasia underwent abdominal hysterectomy under general anesthesia supplemented with regional analgesia. Since preoperative assessment suggested difficult airway, regional anesthesia was planned which was not successful due to spinal abnormality. Subsequently general anesthesia was established with Laryngeal Mask Airway (LMA) with controlled ventilation without any complication.

Keywords: Achondroplasia, Laryngeal mask airway

INTRODUCTION

Achondroplasia is the most common cause of dwarfism which results from abnormal cartilage formation at epiphyseal growth plates. These patients present with short stature, short trunk, and disproportionate development. Achondroplasia is transmitted by an autosomal dominant gene.[1,2] These patients present several problems for both general and regional anesthesia. There are reports describing regional anesthesia in such patients. This report describes the anesthesia management of such patient for abdominal hysterectomy under general anesthesia with LMA supplemented with regional analgesia. We failed to find reports of general anesthesia with Laryngeal Mask Airway (LMA) in such patients.

CASE REPORT

A 34 year old achondroplastic dwarf female was admitted for abdominal hysterectomy .She was 105 cm in height and weighed 24 kg. She had short neck, limited head extension, buck teeth and mallampati grade II airway. Examination of her spine showed severe kyphoscoliosis in thoracolumbar area. Her blood investigations were within normal limit. Her pulmonary function tests revealed FVC 0.85L (%predicted 135), FEV 10.67 L (%predicted 169), FEV1/FVC(%) 82.45 (%predicted 119), MVV 59.56 L/min which is suggestive of mild restrictive pulmonary function. Radiological examination of cervical spine and thoracolumbar spine revealed kyphoscoliosis. Anticipating difficult laryngoscopy the difficult airway cart was kept ready in the operation theatre which included small size endotracheal tubes, bougie, LMA and tracheostomy kit.

Ringer lactate solution was started after securing the intravenous access. Spinal anesthesia was attempted in the sitting position as the patient was not comfortable in supine position. After multiple unsuccessful attempts to find the subarachnoid space, it was decided to take help of fluoroscopic guidance which was unsuccessful.

Subsequently it was decided to proceed with general anaesthesia. The patient was made supine after placing support under her shoulders & head to fill the gap due to kyphoscoliosis. The patient was administered inj. ranitidine 50 mg & ondensetron 4 mg along with glycopyrolate 0.2 mg by IV route. General anaesthesia was induced with inj. propofol 80 mg IV followed by inj. suxamethonium 25 mg IV. Classic LMA No.3 was inserted at first attempt and controlled ventilation was maintained with 60% nitrous oxide, sevoflurane & oxygen. A bolus dose of suxamethonium 10 mg intermitantly provided muscle relaxation. Subsequently the patient was turned to right lateral position and epidural anaesthesia was attempted at L2-L3 level. Epidural catheter no 18G was passed without any difficulty. Inj. tramadol 25 mg diluted to 6 ml was given after the test dose. Vital parameters remained stable. After the surgery when suxamethonium, sevoflurane & nitrouse oxide were discontinued, spontaneous ventilation resumed and LMA was removed after deflating the cuff. The post-operative course was uneventful. Patient was discharged on seventh postoperative day. Patient received inj. tramadol 25 mg eight hourly through epidural rout for next 24 hours for pain relief. The catheter was removed after 24 hours. No neurological deficit was noted.

DISCUSSION

We would like to stress that these patients have several derangements which have potential for complications during administration of anaesthesia.

There are reports describing the achondroplastic patients with classical symptoms and sign of airway obstruction but no difficulty encountered.[3,4] These patients have respiratory problems that may complicate general anesthesia, particularly restrictive lung disease due to thoracic kyphoscoliosis. There is ventilation perfusion mismatch due to decreased FRC and increased closing volume leading to atelectasis. We had done thorough evaluation of pulmonary function which revealed mild restrictive disease.

Several anatomical abnormalites in such patients complicate regional anaesthesia techniques. Kyphoscoliosis associated with narrowing of the vertebral canal, reduced distance between the pedicals and osteophyte formation can produce technical difficulty for spinal anaesthesia.[5,6] In spinal stenosis, the spinal cord is compressed by a spinal canal that is narrowed by abnormally shaped vertebrae. This leads to narrowing of the subarachnoid space and epidural spaces. Spinal stenosis may impair cerebrospinal fluid flow such that identification of dural puncture is more difficult. In this patient repeated dry tap was obtained on attempting lumbar puncture.[7,8]

Detailed preoperative evaluation of the airway should be done. The base of the skull is short and angulated yielding limited extension and making endotracheal intubation potentially difficult, problems that are major causes of perioperative mortality and morbidity. Laryngeal exposure via direct visualization may be difficult in dwarfs with cervical kyphosis and neck flexion should be avoided which can aggravate cord compression in patients with atlantoaxial instability or foramen magnum stenosis.[9] In this patient we had anticipated difficult airway for which difficult airway cart was kept. LMA is an excellent device that can be placed relatively easily to provide a means of oxygenating a patient. It has become the choice for oxygenating a patient when endotracheal intubation has failed.[10] It does not need any ancillary equipment for its insertion. It is safe for elective surgeries where airway pressure does not exceed 20 cm of water. It has been successfully used in difficult airway due to fixed neck or poor mobility of the neck. Difficult Airway Society guidelines which are universally accepted, have recommended the role of LMA in difficult situations.[11,12] We failed to find reports of general anaesthesia with LMA in such patients in anesthesia database.

In conclusion, the risks of both regional and general anesthesia in achondroplastic patients are known. This report describes the successful anesthesia management using LMA for general anesthesia supplemented with epidural analgesia in such patient for abdominal hysterectomy. This case indicates that there could be difficulty in regional anesthesia in such patients as described in other reports. However, general anesthesia with LMA can be safe alternative.

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