ANAESTHETIC MANAGEMMENT OF PREGNANT HYPOCHONDROPLASTIC PATIENT FOR CAESAREAN SECTION - A CASE REPORT

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Summary

We are presenting the anaesthetic management of a hypochondroplastic dwarf pregnant patient with twin pregnancy and severe PIH for elective caesarean section under lumbar epidural anaesthesia. The block could be achieved successfully with six ml of bupivacaine with minimal haemodynamic effects. Sensory block reached upto T4 within 12 minutes with 6 ml. of 0.5 % bupivacaine, with stable haemodynamics and excellent intraoperative conditions. Twin neonates (one male and one female) were extracted successfully with normal APGAR scores. The different systemic and anaesthetic considerations and the anaesthetic management are discussed.

Key Words

Hypochondroplasia, Pregnancy/twin pregnancy, PIH, caesarean section, epidural anaesthesia, bupivacaine, APGAR

Case report:

A 35 year pregnant lady was admitted to the department of obstetrics and gynaecology at Vijayanagara Institute of Medical Sciences (VIMS), Bellary with history of 34 weeks of amenorrhoea and breathlessness for 2 days. She was diagnosed to have twin pregnancy with severe PIH. Her past history revealed that in her married life of 10 years, she conceived 4 times, including the present one. The first one resulted in still birth, second in a full term normal home delivary and the third in IUD (full term hospital delivery). During the present pregnancy, patient has not undergone any antenatal check-ups and not received any immunization. By history, first and second trimesters were uneventful but she developed breathlessness of grade III in the 34th week with no orthopnoea / PND. Her blood pressure was recorded on admission as 200/140 mm of Hg and heart rate of 90/min. There was no history of PIH in the previous pregnancies. She did not give history of previous episodes of breathlessness, chest pain, headache, blurring of vision, swelling of feet, pain abdomen or decrease in urine output. There was no history of sleep apnoea. There was no history of convulsions, jaundice, diabetes-mellitus, tuberculosis, bronchial asthma, allergy or surgery. Family history was not significant. On admission, she was put on tablet nifedepine 10mg thrice a day and tablet alprazolam 0.25mg twice a day for her hypertension. Her daily recordings of BP and HR showed control to the extent of 160/110 mm of Hg and 100 bpm respectively. No drugs were added for further control.

At the time of pre-anaesthetic examination (on the fourth day after admission), BP was 160/112 mm Hg and pulse 102/min with regular rhythm. There was moderate pallor with no jaundice or oedema. Airway was class II to III (Samsoon and Young classification), rule of 123 showed normal values, flexion on the neck was around 90 degrees with slight restricted extension at the atlanto occipital joint. She had almost full set of teeth with a relatively large mandible and large tongue. With the patient awake and with explanation, we used 15 % lignocaine spray and performed gentle direct laryngoscopy and could visualize the glottic opening. She measured 136 cm in

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height and 38 kg in weight, had low set ears and thoracic kyphoscoliosis, lumbar lordosis and bowing tibia of both the legs. Bridge of the nose was depressed with a relatively large cranial vault.

Examination of the respiratory system revealed a slight shift of trachea to the left equal but generally reduced air entry bilaterally and chest expansion of only 1 cm. Cardiovascular system was normal, CNS examination revealed a typical gait of pregnancy lordosis and the waddling associated with kypho-scolosis and normal muscle bulk and power.

Investigations revealed haemoglobin of 8.5 mg % but other parameters including blood counts, Liver and renal functions and coagulation profile were normal. Pulmonary function testing could not be performed due to technical reasons. Ultrasound examination of abdomen indicated twin pregnancy of 32-34 weeks gestation with oligohydramnios. Since the clinical examination of airway was encouraging, and with the subject being a pregnant patient, we did not subject the patient to any X ray (airway / chest / spine). ECG showed sporadic, insignificant ventricular ectopics. Echocardiograph showed dilated dyskinetic left ventricle, reduced left ventricular function with an ejection fraction of 45 % and no evidence of pulmonary hypertension.

Elective lower segment cesaerean section (LSCS) was planned on the 5th day of admission. Since 1. patient had been on antihypertensive medication already, 2.regional anaesthesia is generally preferred in pregnancy (in the absence of contra indications) with respect to maternal and foetal / neonatal safety 3. spinal anaesthesia is likely to cause erratic spread in the presence of hypochondroplasia/ kyphoscoliosis, we planned lumbar epidural anaesthesia in titrated doses in this patient.

Patient was explained about the process of epidural anaesthesia and risks and benefits to her vis-à-vis the neonate and consent was obtained the previous day. The patient was advised to continue tab.nifedepine on the morning of surgery. Oral ranitidine 150 mg. and alprazolam 0.5 mg were advised the previous night.

Patient was shifted to operation theatre in left lateral position and intravenous line could be secured easily in right forearm with a 18# SWG cannula. Ringer Lactate 500 ml. was given as preload. Patient was given injection ranitidine 50 mg. IV and inj Ondansetron 8 mg. IV as premedicants. Routine monitors including pulseoximetry, NIBP and ECG were connected and basal parameters were obtained. The BP was 170 / 116 mm Hg and pulse 110 / min., with normal ECG. Since the patient was otherwise normal with stable respiratory and foetal parameters , it was decided to go ahead with institution of epidural anaesthesia. Anaesthetic drill was performed ; Inj. Ephedrine, Inj. Atropine, Inj. Thiopentone and Inj.Succinyl choline were kept ready. Examination of spine in the left lateral position revealed very narrow spaces in the lumbar region; Epidural catheterization (no.16 G) was hence attempted in the L3-L4 interspace in sitting position and was successful, at a depth of 3.3 cm. Patient was returned to supine position with a 15^0 left tilt, but the table flat end on. Patient could be positioned comfortably on the back and the bowed legs needed support of pillows. A test dose of 3 ml. of 0.5 % Bupivacaine was administered and intravascular and intrathecal placement of the drug were ruled out (over 5 mins.); patient developed sensory block to this dose upto the level of L1-2 by 6th minute. Additional 3 ml. of Bupivacaine 0.5 % was given through the catheter at this point and after about 6-7 minutes, the level of sensory block reached T4 on the left side and T6 on the right side. Grade III motor blockade (Bromage Scale) was reached after 15 minutes.

Oxygen was administered by Magills circuit and obstetrician asked to start the procedure. A Pfannensteil incision was used and skin incision delivary (I D) time was 6 minutes and uterine incision delivary (U D) time was 90 seconds. Twin babies, one female and one male were extracted one minute apart and both the neonates were healthy with good APGAR scores of 8-10 at first and fifth minutes. As the patient appeared anxious, after extraction, inj. midazolam 1 mg. was given IV and oxygenation continued for a further 10 minutes. Patient was comfortable subsequently till the end of procedure. Inj Oxytocin 10 units was infused with Ringer lactate (750 ml) totally. Since there was no significant uterine bleeding and since patient was stable, no colloids / blood / blood products administration was deemed necessary inspite of preexisting anaemia. At no point did the BP fall and was stable between 160-170 mm Hg Systolic and 106 – 112 mm Hg. Diastolic throughout.. In fact there was a single episode of hypertension with a BP of 180/ 120 mm Hg., which stabilized with no treatment..

Patient was shifted out of the OT after exactly 70 minutes of epidural injection.The sensory block was at the level of T8 and motor block still grade III at this time. The first demand for analgesia in the post operative period was 35 minutes later and patient was given inj. Bupivacaine 0.25% with inj. Tramadol 25 mg (to a total volume of 4 ml.) and for the next two days, the same dose was given three times / day. Inj. Ondansetron 4mg. was given IV (B.D). as prophylactic antiemetic.

Both the neonates were normal and the mother was decatheterised on 3rd day and discharged on eighth post operative day without any complications.

Discussion :

Dwarfism is defined as failure to achieve 148 cm. of height by adulthood and more than 100 varieties of dwarfism are described[1]. Hypochondroplasia and achondroplasia are more common in females and the overall incidence is 1.5 in 10,000 live births. It is transmitted as an autosomal dominant gene but only 20 % are familial. The predicted height for an achondroplastic male is 132 cm. and female is 122 cm[2]. Hypochondroplastic individuals are generally taller than achondroplastics and the skull need not be involved. Otherwise, they can have similar clinical features. Basically there is decrease in rate of endochondral ossification formation but normal periosteal bone formation leads to short tubular bones. Mental and skeletal muscular systems are normal with normal life expectancy. Genu varum and kyphoscoliosis are common and the latter may, over the years lead to cor pulmonale / pulmonary hypertension. Premature skull bone fusion with shortening and stenosis of foramen magnum are common in achondroplasia (See below)

Anaesthetic Considerations in Achondoplastic dwarfs*

Upper airway obstruction
Difficult exposure of glottic opening
Kyphoscoliosis ,Restrictive Lung Disease
Cor Pulmonale, Pulmonary Hypertension
Obstructive sleep apnoea, Central Sleep apnoea
Compressive spinal cord and nerve root syndromes
Foramen magnum stenosis
Altered thermal regulation (Hyperthermia)

***** Adopted from Anaesthesia and Co existing Disease[2]

Situations where general anaesthesia is needed should prompt the anaesthesiologist to look at the possible difficulties and manage the patient carefully. History of sleep apnoea may predispose to development of upper airway obstruction after sedation or induction of general anaesthesia. Difficult mask ventilation because of bad fit and difficulty in maintenance of patent upper airway are commonly encountered.

Fertility rates are generally low in these patients. Female patients with pregnancy will be subjected to caesarean section because of small contracted pelvis and infants have near normal birth weights. Presence of PIH (as in the presnt patient) with associated oedema in the upper airway may add on to difficulty in laryngoscopy and intubation. Need to avoid general anaesthesia in pregnant patients (need for awake patient, risk of regurgitation aspiration, foetal considerations,etc.,) and since the caesarean section was on an elective basis, we could plan an epidural technique in advance. However, there is always a risk of an emergency surgery under general anaesthesia in obstetric patients but our preoperative assessment in regard to airway was encouraging. Kallman and colleagues reported successful general anaesthesia for emergency caesarean section in a 98 cm achondroplastic dwarf [3].

Achondro- / Hypochondroplastic patients are in general more anxious compared to others. Since our patient already had PIH, we continued the same on the day of surgery and additionally, midazolam 1 mg IV was given after extraction of foetus.

Technical difficulties may be encountered during epidural and spinal anaesthesia because of kyphoscoliosis and narrow epidural and spinal spaces. No evidence based dosage guidelines are available but it is advisable to titrate an epidural dose to effect, to reduce complications. One of the possible contra indications for regional blocks is the risk of neurological sequelae of spinal abnormality being attributed to the anaesthetic procedure. Since the

patient in the present study did not have any neurological deficits and there were instances of successful management of pregnant achondroplastic dwarfs in the past[3,4,5], we decided to administer epidural anaesthesia. Also, narrowing of the vertebral canal associated with the condition as also the pregnancy changes can lead to higher spread of local anaesthetic in the epidural space. Hence, spinal anaesthesia may actually be more dangerous in this regard with risk of extensive spread.

Kyphoscoliosis and bowed legs made a lateral position for sighting the epidural space difficult in our patient and hence sitting position was used and was successful in first attempt.

Aortocaval compression of pregnancy may be complicated because of kyphoscoliosis with resultant exaggerated hemodynamics. With maintainence of LUD, we did not encounter significant problems and both BP and HR were maintained at preoperative levels.

Wardell and Frame used 5 ml. of 0.5 % bupivacaine (3 ml as test dose and additional 2 ml titrated) in a 111 cm., 46 kg achondroplastic pregnant patient witht marked thoracolumbar kyphoscoliosis, for a block which reached T6 level[5].

Morrow and Black[6], in a 120 cm. achondroplastic pregnant patient, used lignocaine with adrenaline and fentanyl 37.5 μgm, to a total volume of 12 ml. given over 25 minutes to obtain a block of T3-4; they inserted epidural catheter in sitting position, administered a test dose of 1.5 ml. of lignocaine (2%) with adrenaline and a further dose of 6 ml. Patient was then turned supine and additional doses of 3 ml. each of the same drug were given in left lateral and right lateral positions, before being turned " supine with LL tilt". The dose appears larger compared other studies. They had to administer 15mg. of ephedrine for correction of hypotension.

Conclusion :

Pregnant patients with dwarfism can be successfully managed for caesarean section using titrated approach to lumbar epidural anaesthesia. Considerations of kyphoscoliosis and altered epidural and spinal drug distribution and of obstetrics should be borne in mind and this approach can lead to good maternal and foetal / neonatal outcome.

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