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Anaesthetic management of neonatal primary hyperparathyroidism

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Abstract

Neonatal primary hyperparathyroidism (NPHP) is a rare disease that presents in the first 6 months of life. It is almost invariably fatal unless a prompt diagnosis is made and urgent surgical intervention is instituted. Patients with hyperparathyroidism require special preoperative considerations. They require proper work-up from cardiac, renal and neurological point of view, correction of intravenous volume and electrolytes irregularities.

We are presenting a case of an infant who started to have cyanosis and lethargy at 21st day of life. Initial diagnosis of pneumonia was made but later on diagnosed as hyperparathyroidism. He was planned for parathyroidectomy under general anaesthesia. He was induced with sevoflurane followed by fentanyl and atracurium and intubated with size 3.5 mm endotracheal tube (ETT). Later, he was maintained with isoflurane and O2/NO2. He was successfully extubated at the end of operation and was shifted to intensive care unit (ICU) for close monitoring.

Keywords: Hyperparathyroidism, Anaesthetic management, Hypercalcemia.

Introduction

Neonatal primary hyperparathyroidism (NPHP) is a rare but life threatening presentation in which neonates have marked hypercalcaemia, very high levels of parathyroid hormone, hypotonia and respiratory distress.[1] About 50 neonates with NPHP have been reported and it has been suggested as a possible contributor to a small number of cases of the Sudden infant death syndrome (SIDS) as well.[2]

The etiology is genetic and results from inactivating mutation of calcium sensing receptor gene (CaSR), in which the inhibitory effect of extracellular calcium on the secretion of PTH by chief cell is absent.[3] Most reported cases have been of term infants,[4] there has been 1 report of an affected premature infant.[5]

In NPHP, medical therapy is inadequate and surgical modality should be considered. Medical therapy without surgical intervention has a high mortality rate (70-87 %) and results in severe long term complications in the survivors.[6] Parathyroidectomy with auto transplantation provides effective treatment for primary and secondary hyperparathyroidism with a predictable response of symptoms related to hypercalcaemia and elevated parathyroid hormone.[7] This intervention must be undertaken immediately to avoid death or irreversible long term complications such as nephrocalcinosis, cardiac abnormalities, bone resorption or central nervous system alterations.[6]

Case History

This is a case of a 21 days old infant who was brought initially to local district hospital with cyanosis of face, palms and soles, lethargy and irritability. An initial diagnosis of pneumonia was made and treatment started but symptoms did not improve much. He was referred to us in a tertiary hospital of Karachi Aga Khan University almost 15 days after starting treatment. On initial examination, his weight was 3.8 kilograms. He was vitally stable with heart rate of 125 beats per minute, blood pressure 98/58 mmHg, respiratory rate 36 per minute. His breathing pattern was normal with oxygen saturation maintained at 99% without any need of supplemental oxygen. The child was active and there was no neurological deficit. His sucking was strong and sustained. Chest auscultation revealed occasional fine crepts at right base of lung but chest X-ray findings were normal. Blood investigations showed high serum calcium (15.5mg/dl), Parathyroid hormone (PTH) (1690 pg/ml), alkaline phosphatase and low urinary calcium and phosphate. Other investigations included haemoglobin 13.2 g/dl, sodium 127 mmol/L, potassium 3.8 mmol/L, chloride 97 mmol/L, bicarbonate 19 mmol/L, BUN 5 mg/dL and creatinine 0.4 mg/dL. The rest of endocrinological profile was found to be normal. Ultrasound Kidney Ureter and Bladder (KUB) showed bilateral nephrocalcinosis. Hence, hyperparathyroidism was diagnosed and total parathyroidectomy was planned.

Preoperatively, intravenous 1/5 dextrose saline was started at the rate of 25ml/hour along with few doses of intravenous furosemide in the ward to correct hypercalcemia. Echocardiogram was done which was found normal. General anaesthesia was planned and inhalational induction was done with sevoflurane accompanied by injection atracurium 2 milligrams and fentanyl 2 micrograms. Patient was intubated with size 3.5 PVC endotracheal tube. Anaesthesia was maintained with O2/NO2 in 40/60% with isoflurane and additional 2 micrograms fentanyl and 1 miligram atracurium were given intravenously. Parathyroidectomy was done and part of gland was implanted in right brachioradialis muscle. Intraoperatively, patient received intravenous 1/2 dextrose saline at a rate of 40 ml/hr. Patient remained haemodynamically stable and temperature was maintained between 35.2 to 35.5°C

throughout the procedure. At the end of the procedure, patient was reversed with injection Neo-atropine and was extubated when he started opening his eyes and flexing arms and hips.

On shifting to recovery room, his heart rate was 145 beats per minute, blood pressure 65/30 mmHg and oxygen saturation 99 %. There were two very brief episodes of apnea in recovery room so he was planned to be shifted to intensive care unit for apnea monitoring and serial calcium levels. He received two additional intravenous doses of 0.2 micrograms Fentanyl for postoperative pain and was then continued on regular paracetamol suppositories. He remained stable in intensive care unit and serum calcium levels gradually dropped to 10.1 mg/dl while low serum potassium was replaced. His intravenous fluid was changed back to 1/5 dextrose saline at a rate of 15ml/hour in intensive care unit. He also received alpha-calcidol and Ca-gluconate postoperatively. Later on, he was discharged home on 6th postoperative day on oral calcium-gluconate.

Discussion

Two recent changes have radically reduced anaesthesia involvement in the care of patients with primary hyperparathyroidism. One is the role of the calcimimetics especially in older individuals and the other is to use minimally invasive approaches after imaging procedures with just local anaesthesia or a cervical plexus block.[8]

Patients with moderate hypercalcemia who have normal renal and cardiovascular function present no special preoperative problems. The ECG can be examined preoperatively and intraoperatively for shortened PR or QT intervals. As severe hypercalcaemia can result in hypovolaemia, so normal intravascular volume and electrolyte status should be restored preoperatively as was treated in our case. Additionally, proper work up from cardiac, renal and neurologic point of view is very important. In emergency situations, vigorous expansion of intravascular volume usually reduces serum calcium to a safe level (< 14 mg/dl); administration of furosemide is also often helpful in these situations. Other measures to decrease reabsorption of bone include the bisphosphonates pamidronate and zoledronate, salmon calcitonin, and plicamycin.[9]

General anaesthesia is most commonly used, but cervical plexus block and local anaesthesia with hypnosis have been used successfully in adults.[10] Due to the unpredictable response to neuromuscular blocking drugs in the hypercalcemic patient, a conservative approach to muscle paralysis makes sense. There is an increased requirement for vecuronium and probably all nondepolarizing muscle relaxants during onset of neuromuscular blockade but we used 2 miligram (0.5 miligram per kilogram) of injection atracurium which did well in our case. Acidosis decreases calcium binding to albumin thus increasing the levels of ionized calcium, which can cause life threatening hypercalcaemia, hence it is important to maintain normocarbia.[11] Careful positioning of the osteopenic patient is also necessary to avoid pathologic bone fractures.[12]

Postoperative complications include recurrent laryngeal nerve injury, bleeding and transient or complete hypoparathyroidism. Hence, there is a requirement of measurement of serial serum calcium levels which was done in this case. Unilateral recurrent laryngeal nerve injury is characterized by hoarseness and usually requires no intervention. Bilateral recurrent laryngeal nerve injury is a rare complication, producing aphonia and requiring immediate tracheal intubation. Patients with significant preoperative bone disease may have hypocalcaemia after removal of the PTH-secreting glands. Thus, serum calcium, magnesium and phosphorus levels should be closely monitored until stable.[12]

Conclusion

Neonatal primary hyperparathyroidism is a very rare condition which requires special considerations in terms of cardiac, neurological and renal systems irregularities. These patients need cautious administration of neuromuscular agents and intravenous fluids and electrolytes intraoperatively. Postoperative close monitoring of vitals and serial serum calcium levels are also very important.

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