Letters to Editor

Congenital adrenal hyperplasia due to 17-alpha hydroxylase deficiency with hypertensive encephalopathy, hypoglycemic seizures and adrenal insufficiency

Madam,

We report 5-year-old female child with both congenital adrenal hyperplasia and adrenal insufficiency, posted for squint correction of both eyes under general anesthesia.

Congenital adrenal hyperplasia due to 17-alpha hydroxylase deficiency was diagnosed at the age of 3 months when presented with recurrent vomiting and diarrhea and was managed on steroids. Patient had developmental delay, dysarthria, brisk deep tendon reflexes, stunted growth and hyper pigmentation in the skin crease, nape of the neck. Patient developed hypertensive encephalopathy and hypoglycemic seizures at 9 months and 4 years of age respectively due to stoppage of steroid for 3-4 days by the parents indicating adrenal insufficiency.

Hemogram, liver, and renal profile were normal. Serum 17-hydroxy progesterone, plasma renin activity and serum electrolytes were normal. Serum dehydroepiandrosterone sulfate and serum cortisol were low. Height velocity was 4.5
cm/year (<5 percentile) and bone age of 4 years. Ultrasound of abdomen and pelvis was normal. Magnetic resonance imaging brain showed bilateral capsulothalamic multifocal chronic lacunar infarcts. Patient was on oral fludrocortisone and hydrocortisone.

With all standard fasting protocols, patient was shifted to the operation theatre with the parent. Morning dose of steroid was continued. Additional injection hydrocortisone 50 mg/kg was given intramuscularly half an hour before shifting to theater. All parameters such as electrocardiography, noninvasive blood pressure, pulse oximeter and temperature probes were applied. Anesthesia was induced with sevoflurane 8 vol% in 100% oxygen. Intravenous line was secured. Glycopyrrolate 0.08 mg/kg, fentanyl 1 mcg/kg was given intravenously. Sevoflurane was continued at 4 vol% for 3 min. Intubation was done with uncuffed endotracheal tube no 5.5. Anesthesia was maintained with oxygen, nitrous oxide (1:1) and sevoflurane 1.5 vol% in low flow closed circuit. Intravenous fluid was maintained with Isolyte-P. All vital parameters and EtCO$_2$ were monitored continuously. Blood glucose was measured hourly. Hydrocortisone 40 mg, ondasetron 2 mg, pantoprezole 10 mg was given intravenously. The procedure took 150 min. Extubation was done in fully awake condition. Patient was shifted to pediatric intensive for 24 h. Injection hydrocortisone was given 4 hourly in a tapering manner and injection ondasetron was continued in the postoperative period. Analgesia was maintained with topical and systemic analgesics. Oral fludrocortisones and hydrocortisone were started 4 h after the surgery. Patient was discharged on day 3.

An adrenal crisis often occurs if the body is subjected to stress, such as an accident, injury, surgery, sudden withdrawal of long term corticosteroid therapy or severe infection; death may quickly follow.[1]

The main anesthetic concern in this case were the effects of long term steroid therapy, response to the stress of surgery and general anesthesia, tendency to go for adrenal crisis and the postoperative outcome.

Cautious monitoring of blood glucose, blood pressure, temperature and neuromuscular activity was done in the perioperative period. Patient was found very sensitive to inhalational anesthetic as induction was done within two breaths of 8 vol% of sevoflurane and blood pressure and heart was dropped <20% of the preoperative value. Hence, depth of anesthesia was maintained (at sevoflurane 1.5 vol%) and monitored cautiously and titrated according to the hemodynamic parameters. Adequate analgesia was given with fentanyl and paracetamol suppository to reduce the pain and stress perioperatively. Sevoflurane was chosen as both induction and maintenance as it is rapid acting hemodynamically stable and rapid awakening. Since squint surgery is known for postoperative nausea and vomiting. Adding this stress to this patient is detrimental. Hence, adequate antiemetic and proton pump inhibitors were given intra and postoperatively. To conclude, apart from increased sensitivity to sevoflurane, the intraoperative and postoperative course was uneventful.

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