Vitamin D resistant rickets: What an anesthesiologist should know

Sir,

A 5-year-old child weighing 10 kg and diagnosed with X-linked hypophosphatemic rickets was scheduled for open orchidopexy. On physical examination, he was normotensive with frontal bossing, genu varum, pigeon chest deformity, widening of the wrist, dental caries, and alopecia totalis [Figure 1]. He was receiving supplemental calcium, phosphorus, and Vitamin D. His investigations revealed serum calcium 8.14 mg/dl (9–10.5 mg/dl), inorganic phosphorus 3.33 mg/dl (2.5–5 mg/dl), ionized calcium 4 mg/dl (4.4–5.4 mg/dl), urine calcium/creatinine ratio 0.05 (<0.3 mg/mg), and alkaline phosphatase 2539 IU/L (20–140 IU/ml).

An intravenous (IV) premedication with midazolam 0.05 mg/kg and fentanyl 2 μg/kg was given intravenously in the holding area. Anesthesia was induced using propofol 2 mg/kg and patient was intubated. The patient was maintained on a mixture of oxygen, air, and sevoflurane. Single shot lumbar epidural analgesia was given at the level of L2–3 as the caudal space was obliterated. Injection bupivacaine 0.25% was given in a total volume of 0.5 ml/kg with injection clonidine 2 μg/kg. Care was taken during positioning of the patient, and the pressure points were padded. Hyperventilation was avoided as the resulting respiratory alkalosis can cause hypocalcemia. After an uneventful procedure, the child was extubated after reversal of neuromuscular blockade and complete recovery of muscle tone and power.

Three hours postoperatively, the patient developed drowsiness, irrelevant talk, and inability to sit and hold the head without support. Empirically calcium gluconate 5 ml diluted to 10 ml was given intravenously under electrocardiogram monitoring. Post operatively, the serum calcium was found to be 6.96 mg/dl.. Inorganic phosphorus level was normal (3.0 mg/dl). The patient was started on IV calcium supplementation 200 mg/kg/day in divided doses for 24 h and then, restarted on oral medications as in the preoperative period.

X-linked hypophosphatemic rickets is a disorder of phosphate reabsorption and calcium metabolism due to mutation in the PHEX gene (phosphate regulating endopeptidase) on chromosome 22.[1] There is renal phosphate wasting and inappropriately low levels of Vitamin D. It is necessary to confirm normal to near-normal calcium and phosphorus levels preoperatively to prevent postoperative neuromuscular weakness. Preoperative normalization of calcium levels was not accomplished in our case as patient was asymptomatic. The postoperative mental status changes could have been due to hypoventilation and hypercarbia resulting from respiratory muscle weakness due to hypocalcemia or a direct result of hypocalcemia.[2,3] A level of ionized calcium <2.5 mg/dl is associated with muscle weakness and needs to be treated preoperatively. Hyperparathyroidism may develop due to oral phosphate administration, and periodic parathormone monitoring may be required. Nephrocalcinosis leading to renal insufficiency can occur in patients being treated with calcium and phosphorus supplements due to hypercalciuria. Monitoring of urine calcium/creatinine ratio and annual renal ultrasound helps in preventing this risk.[4] Care should be taken while positioning since these patients are prone to fractures. Positioning for intubation may be difficult due to craniosynostosis or large head circumference. Regional anesthesia may be technically challenging due to ossification of ligamentum flavum.[5] Hypertension occurring due to chronic hyperparathyroidism may have to be treated preoperatively. A 2D echo may be indicated in the presence of hypertension and clinical signs.

Vitamin D resistant rickets can present challenges for surgery and anesthesia. Correction of preoperative calcium levels and perioperative vigilance can prevent manifestations of hypocalcemia. Bone fragility can pose problems during intubation and regional anesthesia.

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Conflicts of interest
There are no conflicts of interest.

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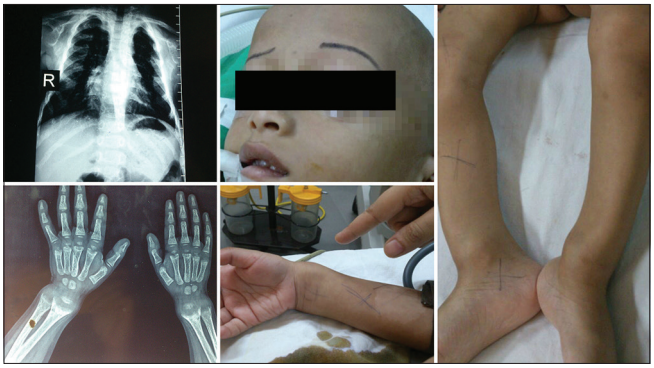

Figure 1: Appearance and radiological picture of patient

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