Anesthetic course of a patient with pineal gland cyst and osteogenesis imperfecta: A rare experience

Sir,

Osteogenesis imperfecta (OI) is an inherited connective tissues disorder of type I collagen formation characterized by brittle bones, hypermobile limbs, kyphoscoliosis, pectus carinatum, short neck, with difficult airway and risk of odonto-axial dislocation during laryngoscopy and intubation.\[^{1,2,3}\] We report anesthetic management of a baby with OI with pineal gland cyst.

A 2½-year-old male child, known case of OI, with magnetic resonance imaging (MRI) documented cyst of the pineal gland with tri-ventricular hydrocephalus [Figure 1] was scheduled for cyst excision. He had suffered multiple fractures of upper and lower limb bones [Figure 2] and was being treated conservatively. Hemogram and serum biochemistry values were within normal range. Chest X-ray revealed thoracic cage deformity with crowding of ribs. MRI findings revealed large cystic area 60 mm × 40 mm × 31 mm in the region of pineal gland causing mass effect on 3rd ventricle with dilatation of lateral ventricles. The patient was carefully placed in supine position. After attaching routine monitors, an intravenous line was secured. After preoxygenation anesthesia was induced with propofol and fentanyl. Airway was secured by endotracheal tube of 4 mm internal diameter after manual in-line stabilization of the neck. Arterial line was secured in right radial artery. Central venous line was placed in right internal jugular vein with the help of ultrasound. Anesthesia was maintained using intermittent IV vecuronium and N₂O in O₂ (1:1) inhalation with variable concentration of sevoflurane. Meticulously patient was kept prone, and pressure areas were padded. A suboccipital craniectomy was done with total excision of the cyst. At the end of surgery, patient was carefully repositioned to supine. Neuromuscular blockade was reversed using neostigmine and glycopyrrolate. After an uneventful extubation, patient was shifted to Intensive Care Unit without any evidence of iatrogenic trauma.

Our case was diagnosed as type IV (A) OI characterized by fragile bone, short stature, and barrel-shaped rib cage. Karabiyik et al.\[^{2}\] used total intravenous anesthesia along with intubating laryngeal mask airway for nephrolithotomy and ureterolithotomy. Malde and Jagtap\[^{3}\] used general anesthesia in case of OI for hysterectomy. These authors reported fracture shaft of the femur while transferring the patient to
the recovery room. Increased intra-operative bleeding may occur in OI\textsuperscript{[4]} due to deficiency of factor VIII and deficient platelet aggregation. Due precautions were taken in the form of availability of adequate blood products. There was no significant blood loss during surgery in our case. Intra-operative hyperthermia is more common in patients with anticholinergic medications and inhalational versus intravenous anesthesia.\textsuperscript{[5]}

In our case, we used sevoflurane as inhalational agent and no hyperthermia was noticed.

We would like to emphasize that in case of OI careful preoperative assessment of the patient is mandatory so as to provide safe anesthesia. When general anesthesia is considered, attention is required for airway management and positioning of the patient. Recognition of potential complications of this disorder is important for anesthesiologist to prevent iatrogenic trauma.

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