Anaesthetic management of a child with adrenoleukodystrophy: A case report

Sir,

Adrenoleukodystrophy (ALD) disorder is characterised by progressive demyelinisation of central nervous system and peripheral adrenal insufficiency resulting from adrenal gland atrophy. ALD is an X-linked disease with six phenotypes, classified according to age of onset, organ involvement and neurological progression rate. The most common form is X-adrenoleukodystrophy of childhood, which is the most severe.\(^1,^2\) Very-long-chain fatty acids (VLCFA) are metabolised by VLC acyl-CoA synthetases in peroxisomes or mitochondrion.\(^2\) Adrenoleukodystrophy gene mutation may impair the peroxisomal import of this synthetase, leading to the accumulation of VLCFAs. It is uncertain whether these long, rigid, acyl fatty acids reduce membrane fluidity, causing an inflammatory response in the nervous system (demyelination) and reduce steroid synthesis in the adrenal glands. Diagnosis is confirmed by abnormally high saturated VLCFAs and C26:C22 ratio in blood or accessible tissues. Magnetic resonance imaging (MRI) of the brain reveals symmetrical hypodense corpus callosum and periventricular white matter.\(^3,^4\)

The patient was 4.5 years old and 18 kg weight female with cerebral type ALD who required general anaesthesia for dental rehabilitation. The written and informed consent was obtained from guardian. The patient was recently diagnosed as a case of cerebral ALD when she presented with left foot-dragging and frequent fall. She was confined to the wheelchair and was demonstrated to have hypotonia. The child showed limited interaction and deafness was noticed.
The preoperative investigations were found to be within normal limits.

Aspiration prophylaxis with injectable ranitidine (1 mg/kg) and ondansetron (0.15 mg/kg) was given. Protection such as a cushion was used at bony prominences to prevent pressure sores. Intravenous hydrocortisone 40 mg was administered prophylactically. The patient was tilted 30° head-up, pre-oxygenated, and cricoid pressure was applied.

Rapid sequence induction was carried out using propofol 2 mg/kg preceded with fentanyl 2 mcg/kg. Muscle relaxation was provided with rocuronium 1 mg/kg. The trachea was nasally intubated with a size 6 North Pole tube. Maintenance of anaesthesia was achieved by sevoflurane. Following the procedure that lasted 120 min, residual neuromuscular blockade was reversed with neostigmine 1 mg and glycopyrrolate 0.08 mg. After the patient demonstrated airway protective reflexes, spontaneous eye opening, and a regular respiratory pattern, the trachea was extubated. Postoperative analgesia was obtained with paracetamol. Recovery was uneventful and the patient was subsequently discharged.

The anaesthesiologist must consider several factors including mental retardation, seizures, hypotonia, gastro-oesophageal reflux, drug history and abnormal adrenocortical function, chronic steroid replacement, and postoperative recovery. Preoperative sedation should be avoided because of the risk of worsening airway obstruction due to hypotonic pharyngeal muscles. Anticonvulsants are continued up to the day of the surgery, and epileptogenic agents should be avoided.

These patients have a higher risk of gastro-oesophageal reflux and pulmonary aspiration. Administration of a histamine-2 antagonist and rapid sequence induction with cricoid pressure is recommended. Another important consideration is patient transfer and positioning. The present patient was confined to a wheelchair, and was susceptible to bone mineral density loss and iatrogenic fracture during transfer. Protection with cushions should be used at bony prominences to prevent pressure sores.

The use of propofol was considered carefully. Commercial propofol is formulated in 10% soybean oil emulsion and does not contain VLCFAs. Unlike thiopentone,[4] propofol[1] provides rapid recovery. Fentanyl was selected for its rapid onset and short half-life.

The trachea was intubated using rapid-onset muscle relaxant with cricoid pressure. We used the nondepolarizing agent rocuronium at a dose of 1 mg/kg for its long action. Succinylcholine is usually used for rapid sequence induction. However, the risk of hyperkalemia in these patients is uncertain with one report of hyperkalemia after succinylcholine used in a patient with multiple sclerosis.[5]

Postoperatively, the patient may be observed for a longer period in recovery room because airway obstruction may worsen due to residual anaesthesia or traumatic oedema. The patient was at a higher risk of gastro-oesophageal reflux and pulmonary aspiration; it is crucial that vomiting is prevented as much as possible hence, use of anti-emetic is recommended.

We have described the successful management of a child with adrenoleukodystrophy. We have mentioned all the investigations, and as a rare case, documentation is always helpful in further evolving management in medicine.

Declaration of patient consent
The authors certify that they have obtained all appropriate parent consent forms. In the form the parent(s) has/have given his/her/their consent for his/her/their child's images and other clinical information to be reported in the journal. The parent(s) understand that their child's names and initials will not be published and due efforts will be made to conceal their child's identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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