Anaesthesia management of a child with Charcot-Marie-Tooth disease for orthopaedic surgery

Sir,

Charcot-Marie-Tooth (CMT) disease is the most common inherited neuromuscular disorder with an incidence of 10-40 in 100,000 worldwide. It is characterised by progressive muscle atrophy, motor and sensory neuropathy. The patterns of genetic inheritance are autosomal dominant (most commonly), X-linked and autosomal recessive. The disorder is typically associated with progressive distal muscle weakness with hyporeflexia or areflexia. There are some concerns about anaesthesia management in these patients such as positioning for surgery and anaesthesia (due to sensory deficits and limb deformities) and using certain drugs, especially neuromuscular blockers. There are also pros and cons about using inhaled anaesthetics (triggering malignant hyperthermia) in these patients. The anaesthesia management in these patients and its impact on the underlying disease is a challenge for the anaesthesiologists.

An 11-year-old girl (weighing 36 kg) and diagnosed with CMT was admitted to our orthopaedic centre for the correction of right foot deformity. Her signs and symptoms included weakness in her legs, ankles and feet, frequent tripping and falling (chief complaint), decreased ability to run and equinovarus. She had a history of surgery for the correction of left foot deformity in another hospital four months ago. She was posted for the correction of right foot deformity [Figure 1]. The routine laboratory tests and brain magnetic resonance imaging were normal. However, the electromyography (EMG) showed severe motor and sensory neuropathy. In the operating room, after performing routine standard monitoring and pre-oxygenation, midazolam 1 mg and fentanyl 1.5 µg/kg were injected intravenously. Anaesthesia was induced with sodium thiopental 5 mg/kg and atracurium 0.5 mg/kg. Anaesthesia was maintained uneventfully with isoflurane 0.5-1.2% in oxygen and a bolus dose of fentanyl (50 µg) and atracurium (6 mg). The right foot deformity was corrected through a 150-min period. At the end of surgery, 0.75 mg of atropine and 1.5 mg of neostigmine were administered intravenously to reverse the neuromuscular block. She was extubated and transferred to the recovery room where she was given 15 mg pethidine intravenously for her pain. Ultimately, she was transferred to the orthopaedic ward after about an hour.

CMT is a progressive hereditary peripheral neurologic disorder, and caused by a specific mutation in myelin genes. It is usually classified into seven types and multiple subtypes. CMT types 1 and 2 are the most common types and CMT type 3 (Dejerine-Sottas disease) is a very severe condition with an early onset of hypotonia during infancy. The patients generally experience slow and progressive distal muscle weakness that may lead to frequent tripping and falls, ankle sprains and talipes equinovarus. Although CMT patients usually have normal life expectancy, it can cause significant disability. Apart from Dejerine-Sottas neuropathy which is sometimes fatal in infancy, the age of onset usually is below 20 years in other types. Some anaesthetic considerations for CMT include surgical positioning, cardiac, respiratory and vocal cord dysfunction, malignant hyperthermia and hyperkalemia associated with the use of succinylcholine and prolonged action of non-depolarised neuromuscular blockers. In patients of advanced age, such abnormal lung functions are exaggerated due to spine deformity (scoliosis or kyphoscoliosis). The respiratory problems can occur with repeated administration of neuromuscular blocking agents; hence neuromuscular monitoring is needed for this condition. Unfortunately, in our hospital, this valuable monitor was not available. Currently, there is no evidence to recommend specific anaesthesia management in these patients. Concerns about the use of nitrous oxide (neurotoxicity from inhibition of methionine synthases), inhalation agents (triggering malignant hyperthermia) and neuromuscular blocking agents have been expressed in case reports.
In our patient, the patient’s parents rejected the use of regional anaesthesia, and there were some restrictions on the use of certain medications and monitors due to our country’s sanctions. Nevertheless, the patient successfully underwent general anaesthesia with isoflurane and atracurium.

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**Conflicts of interest**
There are no conflicts of interest.

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