Anaesthetic Concerns in a Case of Metachromatic Leukodystrophy

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A 22 year old male patient with metachromatic leukodystrophy presented in our hospital for multiple teeth extraction under general anaesthesia. Metachromatic leukodystrophy is a rare congenital neurodegenerative disorder which predominantly affect the corticospinal tract. In this case report anaesthetic concerns and our perioperative management are discussed.

Key words: Metachromatic leukodystrophy, general anaesthesia, anaesthetic considerations, video laryngoscope.

Introduction

Metachromatic leukodystrophy is an autosomal recessive progressive neurodegenerative disease grouped under disorders collectively known as leukodystrophies. Age at onset, clinical manifestations and enzyme deficiency vary for different types of leukodystrophies, so as the anaesthetic implications[1, 2]. We report a patient with metachromatic leukodystrophy who successfully underwent extraction of multiple teeth under general anaesthesia.

Case report

A 22 year old, 50 kg male patient who is a known case of metachromatic leukodystrophy was admitted to our hospital for extraction of multiple teeth under general anaesthesia. Patient had behavioural abnormality and difficulty in walking since 10 years. There was intellectual Disability, drooling of saliva, recurrent gastroesophageal reflux, urinary and bowel incontinence. Patient had convulsions in the past, controlled with oral levetiracetam 500 mg at night. Previous anaesthetic management for MRI was uneventful. Physical examination revealed a heart rate of 76 bpm, blood pressure of 120/80 mm Hg, respiratory rate of 14 breaths/min, oxygen saturation of 99%, bilateral equal air entry with normal vesicular breath sounds and normal body temperature. His airway assessment revealed bilateral patent nostrils, adequate mouth opening and neck movements, thyromental distance < 6 cm, larger tongue with Mallampati class III. Haematological investigations including coagulation parameters, thyroid profile and serum cortisol levels were normal. Serum aryl sulfatase A level was low (81% reduction). Magnetic resonance imaging was suggestive of deep white matter leukodystrophy extending to subcortical white matter with tigroid appearance. Nerve conduction study revealed predominantly demyelinating neuropathy. Electrocardiogram and 2D-echocardiogram were unremarkable except for the presence of grade 1 mitral regurgitation. On the morning of the day of the procedure. pantoprazole 40 mg was given. After confirming adequate fasting status, patient was shifted to the operating room. Standard monitoring was initiated and intravenous access (IV) was secured. Rapid sequence induction and intubation was planned. Both nostrils were prepared using nasal decongestant drops (oxymetazoline 0.05%). After preoxygenation for 3 minutes, IV fentanyl 100 µg was administered. Anaesthesia was induced with IV propofol 100 mg and neuromuscular blockade achieved with IV rocuronium 50 mg. After 90

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seconds well lubricated 7.0 mm ID Ring-Adair-Elwyn (RAE) tracheal tube was passed through the right nostril. Laryngoscopy was performed using C-MAC and nasotracheal intubation was accomplished. Endotracheal tube was fixed after confirming bilateral equal air entry and throat pack was inserted. Nasopharygeal temperature probe was inserted through left nostril. Anaesthesia was maintained with nitrous oxide in 40 % oxygen and titrated concentration of sevoflurane to a MAC of 1.3. Intraoperative period was uneventful. IV paracetamol 1 g was administered as supplementary analgesia. At the end of the procedure which lasted for 5 hours, anaesthetic gases were tapered and residual neuromuscular blockade reversed with IV neostigmine 2.5 mg and glycopyrrolate 0.5 mg. IV ondansetron 6 mg was administered. Temperature probe removed and nasopharyngeal airway was introduced through the same nostril to prevent upper airway obstruction after extubation. Awake extubation was done. Patient was observed in the post anaesthesia recovery room and later shifted to the ward.

Discussion

Leukodystrophy is a group of disorder which primarily affect the white matter of central nervous system. Metachromatic leukodystrophy is a disorder of myelin metabolism characterised by a deficiency of enzyme arylsulfatase A, the gene encoding which is being located on chromosome 22q13.13.[1] Absent or deficiency of ASRA leads to accumulation of cerebroside sulfate within myelin of nerves, causing myelin breakdown. Patients may have early involvement of corticospinal tract resulting in tremors, spastic quadriparesis or paraparesis. Disease is broadly divided into late infantile, juvenile and adult forms depending on the age at onset.[1] Patients with leukodystrophy may present to an anaesthesiologist at different age, both for diagnostic and therapeutic procedure under sedation or general anaesthesia. Anaesthetic technique and intraoperative monitoring should be tailored based on individual needs.[3, 4] Important anaesthetic concerns are difficult airway, risk of aspiration, seizure on multiple drugs causing hepatic enzyme induction, hyperkalaemic response with depolarising muscle relaxants, hypothermia and delayed postanaesthetic recovery. Hepatic enzyme induction alter pharmoacokinetics and pharmacodynamics of many anaesthetic drugs.[5] Difficult airway may be caused by a larger tongue, poor pharyngeal muscle tone, abnormal dentition and higher Mallampati class. Propofol can be safely used to induce these patients. Autonomic dysfuction can cause gastroparesis with increased chances of aspiration during induction. Atrophic denervated muscles are unreliable for neuromuscular monitoring.[6,7] Pressor response to tracheal intubation may be exaggerated and heart rate response to anticholinergic drugs like atropine may be blunted. Autonomic dysfunction can cause profound hypotension under neuraxial block. Pre-existing neurological deficits should be well documented to avert medicolegal issues. A subset of patients may have pulmonary dysfunction and corpulmonale due to progressive scoliosis, who may require spinal deformity correction in the early childhood. Cognitive dysfunction presents a major challenge in communication, assessment and treatment of postoperative pain. Due care should be exercised while positioning the patient to prevent pressure necrosis.[7, 8]

Our patient had recurrent gastroesophageal reflux; to prevent aspiration, a proton pump inhibitor was administered prophylactically and modified rapid sequence induction was performed using rocuronium.[9] Suxamethonium was avoided due to risk of dangerous hyperkalaemic response. Video laryngoscope was used for intubation because of higher Mallampati class. To prevent upper airway obstruction after extubation, we inserted a nasal airway before reversal of the patient. Prokinetic drugs like metoclopramide was avoided because it can cause extrapyramidal side effects. We maintained normocarbia and normothermia intraoperatively. To prevent hypothermia, we used heat and moisture exchanger, forced air warmer and fluid warmer.

To conclude, metachromatic leukodystrophy is primarily a neurodegenerative disease posing a

higher risk of aspiration, autonomic dysfunction, denervation injury, seizure disorder and hypothermia from anaesthetic perspectives. Meticulous preanaesthetic evaluation and execution of plan is required for the safe conduct of anaesthesia.

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