Case Report

Anesthetic management of a case of Sanjad-Sakati syndrome

ABSTRACT

Sanjad-Sakati syndrome is an autosomal recessive genetic disorder first described in Saudi Arabia. Anesthetic management of these patients is challenging due to airway difficulties, electrolyte imbalance, growth and mental retardation, and seizures. The anesthetic management of the syndrome is described in this case report.

Key words: Anesthesia, difficult airway, Sanjad-Sakati syndrome

Introduction

Sanjad-Sakati syndrome (SSS) is a rare genetic disorder first described in the Kingdom of Saudi Arabia in 1988 by Sanjad and Sakati.[1] It has an autosomal recessive pattern of inheritance; the syndrome is characterized by hypoparathyroidism, hypocalcemia, hyperphosphatemia, severe intrauterine and postnatal growth retardation, mental retardation, seizures, craniofacial dysmorphic features (retromicrognathia, abnormal dentition, and dentofacial anomalies), and susceptibility to chest infections.[2,3] These patients undergo frequent surgical procedures for recurrent fractures and dental restorations, they pose anesthetic challenges due to the difficult airway, recurrent respiratory infections, and sensitivity to muscle relaxants.[4,5]

Case Report

Institutional Review Board approval is not required at King Fahd Medical City (Riyadh, Saudi Arabia) for publication of isolated case reports. A 19-year-old Saudi female patient, weighing 9 kg, diagnosed with SSS, is admitted to our hospital for an open reduction and internal fixation of the right femur fracture. She was born prematurely at 32 weeks to nonconsanguineous parents by cesarean section. Postdelivery, she required neonatal intensive care unit (NICU) admission and ventilatory support. Her problem list included hypoparathyroidism, hypocalcemia, nephrocalcinosis, hypothyroidism, growth retardation, seizure disorder, superior mesenteric artery syndrome, recurrent respiratory infections, and constipation. She required frequent NICU admission with mechanical ventilation for respiratory infections and central hypoventilation syndrome diagnosed by polysomnography. Tracheostomy was performed at the age of 16 years, and she was on home ventilation.

The patient underwent correction of the mal-rotated gut at age 15 years. Her home medications included thyroxine, alfacalcidol, hydrochlorothiazide, potassium chloride, magnesium sulfate, and calcium supplements. Preoperative laboratory results were within normal ranges.

The patient came to the operating room with size 5.0 mm cuffed tracheostomy tube. She received midazolam 1 mg intravenous (IV) preoperatively. The patient was attached to the anesthesia machine with a pressure controlled ventilation

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mode. General anesthesia was induced with propofol 50 mg and fentanyl 30 mcg IV. Muscle relaxants were avoided as the patient had small muscle mass and the surgical procedure could be carried out without any difficulty. Anesthesia was maintained with sevoflurane 2%. A right femoral nerve block was performed to provide adequate peri-operative analgesia under ultrasound guidance with 8 ml of 0.25% plain bupivacaine. The surgical procedure was uneventful, and the patient was transferred to pediatric ICU for overnight observation. She was transferred to the surgical ward next morning and was discharged home after 9 days.

Discussion

SSS is a rare genetic disorder first described in the Kingdom of Saudi Arabia in 1988 by Sanjad and Sakati,[1] characterized by congenital hypoparathyroidism, hypocalcemia and hyperphosphatemia, seizures, severe intrauterine and postnatal growth failure, proportional dwarfism with small feet and hands, mental retardation, craniofacial dysmorphism including microcephaly, retromicrognathia, deep-set eyes, depressed nasal bridge, long philtrum, thin lip, and thick earlobe. We present a 19-year-old girl who required anesthetic care for an emergency open reduction and internal fixation of right femur fracture. The perioperative implications of the disorder are reviewed and suggestions for anesthetic management provided.

SSS consists of hypoparathyroidism leading to hypocalcemic tetany, seizures, or both.[1,2] Patients have typical facial features and are mentally subnormal.[3] The patients usually have characteristic features such as the long narrow face, deep-set eyes, beaked nose, large floppy ears, long philtrum, thin upper lip, and micrognathia.[4] They are usually symptomatic in the newborn period with complications of hypocalcemia and investigations reveal a picture of hypoparathyroidism.[1-3] They often come to the operating room for recurrent fractures and dental restorations.[4] These patients pose an anesthetic challenge because of various factors including difficult airway, growth retardation, hypocalcemia, recurrent respiratory infections, and sensitivity to muscle relaxants.[5] Successful anesthetic management of these patients depends on meticulous preoperative assessment and optimization. Thorough preoperative evaluation should be carried out for the potential difficult airway. A difficult direct laryngoscopy and tracheal intubation should be expected, especially in patients with prominent micrognathia. It is advisable to maintain spontaneous respiration until the airway is secured and bilateral lung ventilation confirmed. In our case, the patient was already tracheostomized thus obviating the possibility of difficult airway. Abnormal electrolyte balance is often seen in patients with SSS and needs to be corrected preoperatively. Hypocalcemia, hyperphosphatemia, hypomagnesemia, and hypokalemia are common in these patients.[1,2] Uncorrected hypocalcemia may lead to long QT interval and associated dysrhythmias, hypotension caused by peripheral vasodilation and ventricular dysfunction, seizures, and increased neuromuscular irritability with increased incidence of laryngospasm.[6-8] Chronic use of anticonvulsant drugs may alter the metabolism of certain medications administered during anesthesia, narcotic agents, and anesthetics known to decrease the seizure threshold should be avoided. Respiratory or metabolic acidosis may further decrease ionized calcium by increasing protein binding, and thus increase the likelihood of the above-mentioned complications.[7,8] Seizures are common in these patients and should be controlled preoperatively.[1-3] Developmental delay and associated mental retardation can make the perioperative care more challenging.[3] Recurrent respiratory infections are common in these patients.[9] A thorough examination should be performed. Preoperative chest X-ray and pulmonary function tests should be requested if indicated. Any respiratory infection should be treated preoperatively. These patients have a higher incidence of central hypoventilation syndrome, which should be confirmed by polysomnography. Postoperative ICU or high dependency unit admission should be considered in these cases. Affected patients with SSS may be sensitive to nondepolarizing muscle relaxants due to underlying hypocalcemia.[10] It is advisable to avoid the use of muscle relaxants if surgical and anesthetic conditions permit; if it is necessary to administer a nondepolarizing muscle relaxant, then a lower dose should be used, guided by a nerve stimulator. Neuromuscular blockade should be completely reversed with a train of four ratio of ≥ 0.9 to ensure a good postoperative respiratory function. Postoperative pain control should be optimized to prevent respiratory complications and sufficient enough to prevent hyperventilation — hypocalcemia — tetany cycle.

While effective pain control is essential, associated respiratory involvement and abnormal central control of ventilation may predispose these patients to respiratory depression with opioids. As such, adjunctive agents including nonopioid analgesics (paracetamol) and local/regional anesthesia are suggested to limit perioperative opioid requirements.[11]

Conclusion

SSS is a rare genetic syndrome. Potential perioperative concerns include phenotypic features that may make airway management challenging, seizures, hypocalcemia, recurrent respiratory infections, and sensitivity to muscle relaxants. A

proper preoperative optimization, identifying and preparing for potential difficult airway, maintaining intraoperative acid-base and electrolyte balance, complete neuromuscular recovery and optimal pain control are essential for successful anesthetic management of these patients.

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Conflicts of interest
There are no conflicts of interest.

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