Anesthetic Management of a Case of Sanjad-Sakati Syndrome

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ABSTRACT

Sanjad-Sakati syndrome (SSS) is a rare genetic disorder, also known as Hypoparathyroidism-Retardation-Dysmorphism syndrome. It is an autosomal recessive syndrome primarily observed in the Arab region, and it is associated with multiple abnormalities, including dysmorphic features, electrolyte disturbances, hypoparathyroidism, intellectual delays, and seizures.

INTRODUCTION

Sanjad-Sakati syndrome (SSS) was first reported in the Kingdom of Saudi Arabia (KSA) in 1988. It is an autosomal recessive syndrome primarily observed in the Arab region, though some cases have been reported elsewhere. The syndrome is characterized by hypoparathyroidism, hyperphosphatemia, and intellectual and physical growth delays. Because SSS has multiple craniofacial features, it can be highly challenging for anesthesiologists to manage such cases, particularly their airways. As such, this case report aims to highlight some of the knowledge associated with managing patients with SSS and to increase awareness of a rare case with multiple anesthesia considerations.

CASE PRESENTATION

A 15-year-old female was diagnosed with SSS after being scheduled to undergo an examination under anesthesia, in addition to corneal scrapings and the application of chelating agents to both corneas. The patient was also diagnosed with hypothyroidism, gastroesophageal reflux disease (GERD), bilateral nephrocalcinosis, intellectual delays, craniofacial dysmorphic features, and corneal opacities. According to the pre-anesthesia assessment, she was a full-term baby delivered vaginally to non-consanguineous parents; in addition, there was no previous surgical history, and she had a good functional capacity and cardiorespiratory reserve. On examination, the patient weighed 7.8 kg, and her height was 80 cm, giving her the appearance of being younger than her actual age. Her vital signs were stable, she maintained 100% oxygen saturation breathing room air, and she was active and awake.

An airway examination exhibited a very short thyromental distance estimated to be less than 3 cm, limiting the mouth opening to Mallampati (MP) Class II and signifying micrognathia with good neck movement and poor dental

hygiene. She had normal cardiorespiratory examination results, and additional measurements, including thyroid function, complete blood count (CBC), and electrolyte count, were within the normal limits.

The patient was admitted one day prior to her surgery. When she arrived to the theatre, she had a 24-gauge IV line and an American Society of Anesthesiologists (ASA) monitor attached, which collected noninvasive blood pressure, pulse oximetry, temperature, and electrocardiographic (ECG) data. The room was warmed to 26oC, and a Difficult Airway Cart and video laryngoscope with different blade sizes were checked and prepared. Anesthesia was induced by inhalation of 8% sevoflurane in 8 L/min oxygen flow, and 1 mcg/kg fentanyl was given to facilitate intubation, which was achieved using a C- MAC® size 2 blade and a size 4 RAE tube cuffed with a stylet. The end-tidal CO_2 level signified normal bilateral air entry with good chest rising.

The patient received 0.1 mg/kg dexamethasone and 180 ml of ringer lactate, and the procedure lasted 3 h. The patient was noted to have intermittent episodes of apnea during the procedure, but she maintained a normal homodynamic profile. Extubation was achieved successfully while the patient was fully awake, and she was shifted to the post-anesthesia care unit (PACU), where she remained vitally stable with no acute complications; thereafter, she was transferred to the ward and discharged the following day.

DISCUSSION

SSS was first reported by Sanjad and Sakati in 1988[1], caused by a mutation in chromosome 1q42.3 of the tubulin co-factor E gene[2]It is an autosomal recessive disorder, first discovered in the Middle East region but also reported in other areas, including Jordan and Israel[3] and its incidence in the KSA varies from 1 in 40,000 to 1 in 60,000 live births[4].

The syndrome is characterized by metabolic and physical abnormalities; for instance, patients can manifest hypoparathyroidism, hypocalcemia, hyperphosphatemia, hypokalemia, hypomagnesemia, reduced growth hormone production, seizure disorder, and intellectual delays. Phenotypically, patients usually have a short stature and multiple craniofacial features, such as microcephaly, a depressed nasal bridge, micrognathia, deep-set eyes, a long philtrum, and thin lips[5]. Consanguineous marriage is one of the major predisposing risk factors for the condition[6], and affected patients can present for surgeries, such as the repair of recurrent fractures, examination under anesthesia for corneal pathologies, and dental procedures[7]. SSS may present similarly to other genetic conditions, such as DiGeorge Syndrome or Kenny-Caffey syndrome, with no cardiac abnormalities and a normal T cell[8].

Concerning anesthetic management, patients with SSS can pose a clinical dilemma, as they are at a high risk for difficult airways and subsequent difficult intubations; electrolyte disturbances, namely, hypocalcemia; increased sensitivity to muscle relaxation; and aspiration[9]. Patients should be meticulously assessed preoperatively to identify underlying

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medical comorbidities, such as hypoparathyroidism, hypocalcemia, and seizures, as well as the risk of lung infections. An uncorrected calcium level may also be associated with arrhythmia, potentiated neuromuscular blocks, and an elevated risk of laryngospasm ,Further, a careful airway examination is mandated to assess the possibility of a difficult airway in these patients along with Basic investigations, including CBC, electrolyte count, and a chest X- ray, should be conducted, and a pulmonary function test should be requested, if indicated[10].

Patients should be allowed to breathe spontaneously until the airway is secured. In addition, minimizing the use of muscle relaxants or taking them in only small doses if required, as guided by a nerve stimulator, should be considered[10]. Patients with SSS have a heightened risk of central hypoventilation; thus, they should be monitored in the post-operative stage, either in a high dependency unit or pediatric intensive care unit. In addition, the use of multimodal analgesia can efficiently reduce pain and minimize the risk of respiratory depression.

CONCLUSION

SSS is an autosomal recessive syndrome that has the potential to pose an anesthetic dilemma, particularly in the management of difficult airways, the correction of concomitant electrolyte disturbances, and the accurate assessment of the risk of chest infections and seizures. Thus, meticulous pre-operative optimization is required for successful anesthetic management of these patients, along with a multidisciplinary approach for a successful outcome.

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