CASE REPORT

Sugar-coated Sleep: Raising Dental Red Flags in Smith–Magenis Syndrome

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Abstract

Aim and background: Smith–Magenis syndrome (SMS) is a rare condition characterized by abnormalities affecting chromosome 17 or RAI1, leading to physical, developmental, and behavioral challenges. SMS occurs in approximately 1 in 25,000 individuals, presenting complex clinical and dental issues.

Case description: This case report focuses on the dental care of a 3-year-old child diagnosed with SMS, emphasizing a comprehensive treatment plan. The child exhibited typical SMS traits, including sleep disturbances, developmental delays, and behavioral problems. The multidisciplinary team integrated dental interventions with strategies to manage these challenges effectively.

Conclusion: This report contributes to the limited knowledge on managing SMS, highlighting the effectiveness of a multidisciplinary approach in meeting the diverse needs of affected individuals.

Clinical significance: The scarcity of literature on SMS underscores the importance of documenting such rare cases to enhance understanding and tailor interventions. By documenting successful management strategies, clinicians can better support patients with this rare disorder.

Keywords: Case report, Comprehensive health care, Dental care, Developmental disabilities, Genetic disorders, rare, Smith–Magenis syndrome.

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Introduction

Smith–Magenis syndrome (SMS) (OMIM#182290) is a rare developmental disorder that is intricately intertwined with distinct facial features, sleep disturbances, and a spectrum of behavioral challenges. With an estimated global incidence of one in 25,000 individuals, SMS is typically linked to a chromosomal deletion on chromosome 17, resulting in the loss of the RAI1 gene.[1] Beyond its impact on behavior, emotions, and cognitive processes, this syndrome poses unique and complex challenges in dental health management, particularly in the pediatric population. Dental health in individuals with SMS is significantly shaped by their characteristic facial traits, evolving from subtle nuances in early childhood to more pronounced features as they grow. This visual aspect often conceals the underlying dental concerns and requires specialized attention.

The intricate relationship between the behavioral and cognitive characteristics of SMS individuals and their oral health is a crucial dimension. Manifestations of self-injurious behaviors coupled with the potential impact of psychotropic medications and sleep aids underscore the need for an in-depth understanding of the distinctive dental challenges faced by these children. Given that dental problems in this population can be exacerbated by medication formulations or behavioral aspects, a holistic and collaborative approach is imperative for effective dental care. This paper aims to bridge the existing gap in clinical guidelines by presenting a rare case report focusing on the dental treatment of a 3-year-old boy with Smith–Magenis syndrome. This underscores the need for customized dental strategies, regular check-ups, and preventive measures to mitigate the increased risk of dental caries in this population. A collaborative relationship between healthcare professionals and dental professionals is essential for comprehensive care in managing the complex challenges associated with Smith–Magenis syndrome.

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Case Description

A 3-year-old boy with a known history of Smith–Magenis syndrome was referred by his pediatrician to the dental outpatient department with the chief complaint of multiple decayed teeth accompanied by mild pain. The diagnosis of the syndrome was confirmed by whole-exome sequencing conducted at MedGenome Labs Ltd., Bengaluru. Following a comprehensive family history analysis, no hereditary patterns of the disease were identified, and the parents were in a nonconsanguineous marriage. Furthermore, the mother experienced miscarriage before the patient was born, and no other family members exhibited similar conditions (Fig. 1).

The child was conscious, cooperative, well built, well nourished, and weighed 15 kg (33 pounds). The mother mentioned the child's sleep difficulties, a typical behavior known for this syndrome,

and the child was prescribed syrup risperidone 1 mL/L mg (0.3 mL HS) and syrup melatonin 5 mL/3 mg (5 mL) to facilitate a few hours of sleep each night. Pediatric evaluations reported global developmental delay in the child along with observed hyperactivity. Extraoral examination revealed brachycephaly, short stature with central obesity, upslanting palpebral fissure, midface hypoplasia, flat nasal bone, brachydactyly, and polydactyly (Fig. 2). The upper lip was everted and lymphadenopathy was not observed. Facial symmetry was normal and the facial profile was convex. Intraoral examination revealed a complete set of primary dentition, along with severe early childhood caries (ECC) and generalized marginal gingivitis (Fig. 3). No abnormalities were observed in the buccal mucosa, tongue, floor of the mouth, or frenum except for the presence of a high-arched palate. Owing to the child's uncooperative behavior (Frankel definitely negative) during attempts at radiographic evaluation, no radiographs were recorded.

Given the patient's medical condition and the requirement for extensive dental procedures, the patient was admitted to the hospital for treatment under general anesthesia with postparental informed consent. Cardiac evaluation, including electrocardiography (ECG) and 2D echocardiography, revealed indications of a tiny patent foramen ovale and mild tricuspid regurgitation; however, the child was deemed fit for dental treatment under general anesthesia, as confirmed by a pediatric cardiologist. Laboratory evaluations indicated hypochromic microcytic anemia with neutrophilic and eosinophilic leukocytosis and thrombocytosis. The child had a B-negative blood group; consequently, packed red blood cells were reserved before the procedure as a precautionary measure in anticipation of possible

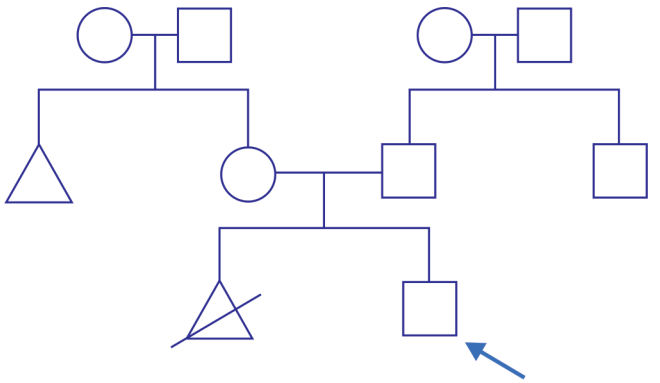

Fig. 1: Pedigree chart of the patient

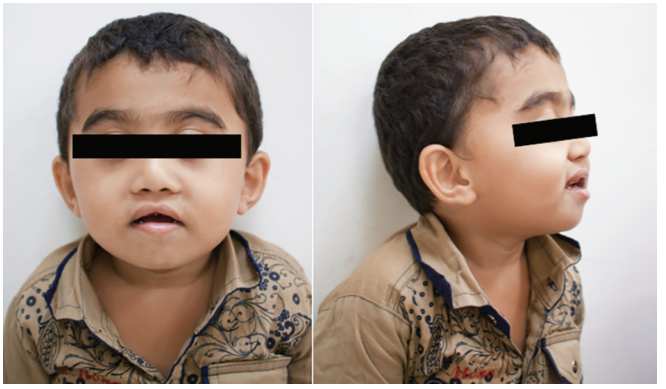

Fig. 2: Extraoral photographs of the patient

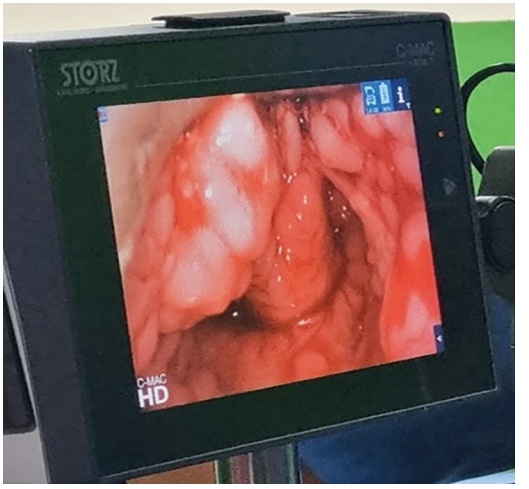

Fig. 4: Enlarged lingual tonsils observed during intubation

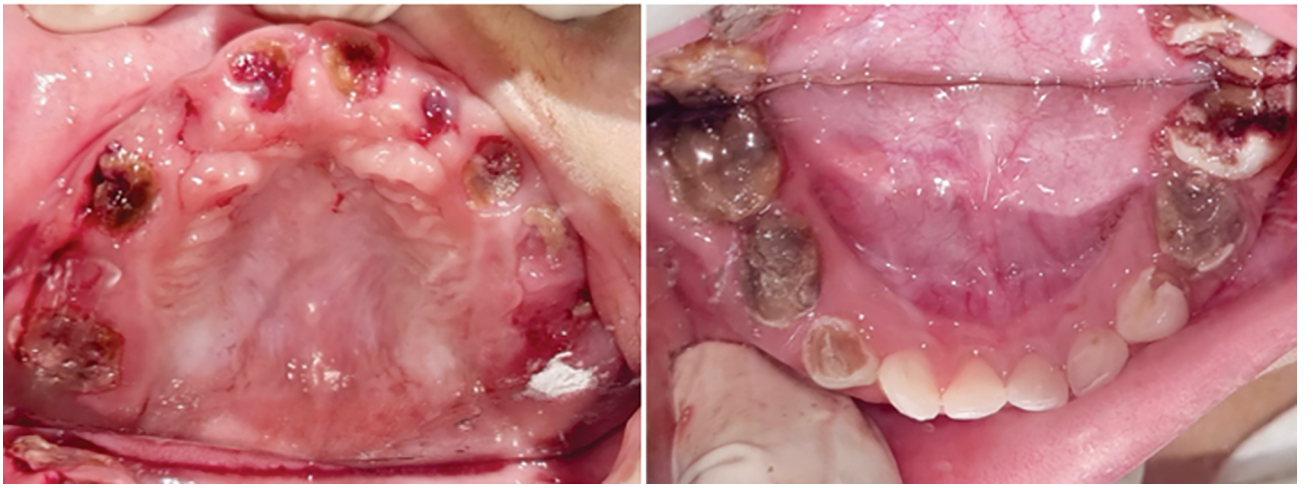

Fig. 3: Intraoral photographs of the patient

blood loss. Nasal intubation proved particularly challenging because of an obstructed airway resembling a suspected enlarged lingual tonsil (Fig. 4). Full mouth rehabilitation involved oral prophylaxis, pulpectomy in the lower left back tooth (tooth #75) using Vitapex (J Morita, Japan), followed by final restoration with a stainless steel crown (3MTM ESPETM Stainless Steel Crowns, 3M, St. Paul, MN, United States). Glass ionomer cement (3 M ESPE Ketac TM Molar, 3 M ESPE Corp., Minnesota, United States) restorations were given on teeth #73 and #83. Extractions were performed on nonrestorable, nonvital teeth #55, 54, 53, 52, 51, 61, 62, 63, 64, 65, 74, 84, and 85. Vicryl sutures (3–0) were placed to approximate the extraction sockets and hemostasis was maintained. In addition, a fluoride varnish (Embrace™ Varnish Pulpdent Corporation, Watertown, CA, United States) was used. After the procedure, intravenous dexamethasone 0.5 mg was administered and the patient was observed in the pediatric intensive care unit (PICU) for 24 hours. The patient exhibited a transient fever of 100°F, which subsided after a few hours of intravenous paracetamol (15 mg/kg/dose). The patient broke the nil-by-mouth restriction (NBM) 4 hours after surgery by consuming clear fluids. Antibiotics and analgesics were prescribed following dental procedures. Postoperative instructions included adhering to a soft diet for 1 week and maintaining oral hygiene.

A 1-month follow-up revealed no complications and a comprehensive approach to subsequent oral care was carefully implemented during this visit. The personalized home oral care routine involved various oral sensory activities, such as gum massage, blowing exercises, and oral motor drills. In discussions with the pediatrician about child medications, efforts were made to explore sugar-free options for medicated syrups to prevent the future occurrence of dental caries. Considering the extraction of multiple teeth, referrals to a dietitian and speech and language pathologist were initiated to reinforce nutrition, and phonetics, respectively.

To address the challenge posed by a child's inability to spit, several recommendations have been suggested, including the use of fluoride-free toothpaste that is safe for minimal consumption, soft or electric toothbrushes, routine tongue scraping, and postmeal mouth rinse facilitated by an irrigation syringe (Fig. 5). During recall visits, the application of topical 5% sodium fluoride with casein phosphopeptide–amorphous calcium phosphate complex (CCP–ACP) remineralization paste on the retained teeth was recommended every 3–6 months. This strategic intervention aimed to progressively improve oral health. Evaluation of plaque and gingival indices before and after implementation of the home care regimen demonstrated a significant improvement after 6 months of follow-up, highlighting an improved oral quality of life.

Fig. 5: Facilitating oral hygiene maintenance using an irrigation syringe

Discussion

Smith–Magenis syndrome (SMS) is a rare and intricate developmental disorder characterized by a range of behavioral, cognitive, and physical challenges. Notable physical features include short stature, scoliosis, obesity, hoarse voice, and abnormalities of the ears and eyes. Distinctive facial features mark individuals with SMS, and one notable hallmark of the syndrome is sleep disturbances that affect both nighttime sleep and daytime drowsiness. Alongside these challenges, common behavioral issues include temper tantrums, aggression, anxiety, impulsiveness, and attention difficulties.[1,2] These difficulties can hinder compliance with at-home oral hygiene measures, consequently increasing the susceptibility to dental caries. Moreover, individuals with SMS often require psychotropic medications and sleep aids, further increasing the risk of dental decay.[3] The complex interplay of physical, behavioral, and medical factors underscores the multifaceted nature of the challenges faced by individuals with Smith–Magenis syndrome. Due to the scarcity of extant literature concerning the reported dental management of Smith–Magneis syndrome, our case report endeavors to elucidate a comprehensive approach. This aims to furnish pertinent insights to augment the knowledge base available to pediatric dentists and contribute to the broader scholarly discourse on this distinctive syndrome.

Children with SMS are predisposed to dental caries, particularly because of the potential sugar content in their sleep medications. Common side effects associated with the use of sleep medications in children include nausea, vomiting, and xerostomia, which further make these children more prone to dental decay. A prudent approach involves the recommendation or formulation of sugar-free sleep medications, along with customized oral hygiene practices and dietary adjustments.[4] Furthermore, upon recording the diet history, it was observed that the child consistently consumed a cariogenic diet. In addition, the child was still breastfeeding. Continued breastfeeding has been correlated with elevated susceptibility to dental caries among children.[5] Hence, the recommendation to start weaning and reduce the consumption of sweet and sticky foods was developed, coupled with the introduction of probiotics.[6,7] This approach aims to mitigate the potential risk factors associated with dental caries and foster optimal oral health for the child.[8,9]

Conclusion

Smith–Magenis syndrome presents distinct challenges in pediatric dental care, requiring collaboration between pediatricians, pediatric dentists, dieticians, speech and language pathologists, and other specialists. The integration of sugar-free sleep medications and prompt referral to pediatric dentists is crucial to mitigate the increased risk of dental caries. Dieticians play an essential role in providing guidance to achieve adequate nutrition, especially after multiple tooth extractions.[10] Tailored strategies, including rigorous oral hygiene, preventive therapy, and continuous dental risk assessment, are essential for managing the oral health of these individuals. Ongoing research and interdisciplinary collaboration are essential to refine dental care protocols for children with Smith–Magenis syndrome.

Clinical Significance

Smith–Magenis syndrome (SMS) does not significantly affect life expectancy, and people with this syndrome can live into adulthood.[1] However, quality of life can be affected by various challenges that require a forward-looking approach to dental rehabilitation. In response to these challenges, oral sensory exercises were recommended, designed not only to stimulate the sensory muscles of the mouth but also to prevent potentially harmful coping mechanisms resulting from regression in daily activities. While the potential use of removable prostheses offers various benefits, including addressing cosmetic concerns and facilitating societal acceptance and adjustment, the child's young age and behavioral immaturity posed challenges to their ability to cooperate with wearing such prostheses in our case. The child will be subjected to ongoing follow-up, and future treatments will be meticulously planned based on evolving requirements and behavioral considerations. This will encompass space management and the systematic monitoring of guided eruption as integral components of the overall treatment strategy.

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