Solitary Median Maxillary Central Incisor: A Case Report with 3-Year Follow-Up and Literature Review

Abstract
A solitary median maxillary central incisor (SMMCI) is a rare anomaly that can occur alone or be associated with other systemic abnormalities. Early diagnosis of SMMCI is crucial as it might indicate the presence of an associated congenital or developmental abnormality. The prevalence of live-born children with SMMCI is determined to be 1:50,000 and is more common among females. The purpose of this paper was to report an unusual case of a 9-year-old girl with SMMCI who had no growth deficiency or any other systemic involvement. Since pediatricians and dentists are the first professionals to evaluate an SMMCI's patient in most cases, it is important that they be aware of the possibility of other related systemic problems that require systemic care. Appropriate treatment, diagnosis, and referral should also include neuropsychiatric evaluation, genetic testing, and craniofacial profile analysis along with multidisciplinary approach.

Keywords: Holoprosencephaly, karyotyping, solitary median maxillary central incisor

Introduction
A solitary median maxillary central incisor (SMMCI) is a rare anomaly that can occur alone or be associated with other systemic abnormalities. The best-known association is with holoprosencephaly (HPE) and other systemic abnormalities like growth deficiency syndromes or association or specific chromosomal abnormalities. The presence of unexplained SMMCI is considered a risk factor even in the absence of any other clinical signs. The purpose of this paper was to report an unusual case of a girl with SMMCI who had no growth deficiency or any other systemic involvement.

The prevalence of live-born children with SMMCI is determined to be 1:50,000 and is more common among females. Early diagnosis of SMMCI is important as it may be a sign of other severe congenital or developmental abnormalities and additional syndromes. All cases of HPE are associated with SMMCI syndrome, but the inverse is not true.

In this case, a 9-year-old female patient was referred due to trauma to the upper front tooth. Intraoral examination revealed a SMMCI. The diagnosis of such an association entails important implications for the patients' physical and mental development, as well as an elevated risk of manifestation in progeny. Therefore, it is essential that such midline defects when observed in dental development, though mild in nature, should be thoroughly investigated. As in most cases reported till date, the dentist is the first professional to evaluate a patient with this condition.

Case Report
A 9-year-old girl visited the department of pedodontics and preventive dentistry with the chief complaint of throbbing pain in the upper front tooth. The patient’s mother gave a history of trauma before 1 year. The tooth had fractured but was asymptomatic. The patient had visited a dentist for a fractured tooth and had a tooth-colored restoration done, which had fractured within a week thereafter. The patient had a throbbing pain in the upper front tooth since 2 days, especially on lying down; hence, she reported to our department. General examination revealed right-hand thumb agenesis with no significant known medical history. Intraoral examination revealed a single central

How to cite this article: Nalawade TM, Mallikarjunam, Sogi HP, Bhat KG, Kumbar VM. Solitary median maxillary central incisor: A case report with 3-year follow-up and literature review. Contemp Clin Dent 2021;12:324-7.
incisor at the maxillary midline having a fractured incisal edge involving enamel and dentin [Figure 1a]. No dental avulsions or tooth extractions in the region of the incisors were reported. In addition, she had maxillary atresia, the philtrum and incisive papilla were indistinct [Figure 1b], and the maxillary frenum was absent. Both parents were of normal stature and were healthy. There was no history of consanguinity or hereditary disease in her family.

Orthopantogram showed a single central incisor at the midline and all other permanent teeth were present except third molars [Figure 1c]. A radiographic investigation by a posteroanterior view [Figure 1d] also confirmed the presence of a single central incisor in the midline and suggested deviated nasal septum toward left and a narrow pyriform aperture. Chromosomal analysis by karyotyping was advised and if required, genetic counseling was planned after the analysis. In addition, other systemic disorders were ruled out and intellectual ability seemed to be average after pediatric consultation. Karyotyping with G-banding of peripheral blood cells suggested no major structural and numerical abnormalities of the chromosomes [Figure 2a and b].

As karyotyping results were normal, the 9-year-old girl turned out to be a case of SMMCI without associated systemic involvement due to major structural and numerical defects of chromosomes. This case may be sporadic because there was no evidence of consanguinity or any other defects in siblings or family members. Root canal treatment followed by full coverage restoration was given in this case [Figure 3a and b]. She was satisfied with the appearance of her teeth and was not aware of her orthodontic problems.

Orthodontic and prosthetic treatment options are possible. As there was generalized spacing in maxillary anteriors and the patient was in mixed dentition, fixed orthodontic treatment would be considered at a later stage. After orthodontic alignment, prosthetic replacement of missing central incisor with a removable partial denture could be done. After relative cessation of growth, i.e., around 18–20 years, fixed partial denture or a single tooth implant could also be considered. Preventive care, orthodontic treatment, and esthetic restorative dentistry are important in long-term dental management.

Hence, she was advised regular follow-up every 6 months and she continues to be followed by pediatrician and genetic consultant. After 3 years of follow-up [Figure 3c], the patient has no complaints regarding the esthetics of her teeth. Furthermore, she has showed progressive growth and development, further ruling out any systemic involvement.

Discussion

The presence of a single central incisor positioned on the midline is a rare occurrence. It may be related to first and foremost premature tooth loss due to trauma,[4] followed by hypodontia or less commonly SMMCI syndrome. It was first reported in 1958 by Scott as an isolated finding,[5] followed by Fulstow in 1968. In 1976, Rapport et al. introduced the term “Monosuperocenroincisivodontic dwarfism.” Among several names such as solitary maxillary central incisor and single maxillary central incisor; SMMCI syndrome has been the accepted term to describe the peculiarly formed symmetric incisor tooth occurring with and without other systemic involvement.[6] It can occur either in the primary or secondary dentition.[7] To accurately diagnose a patient with SMMCI, the characteristic tooth must fulfill certain specifications. The tooth present must exist as the only central incisor tooth located precisely on the mid-palatal raphae and not the mandible. If the tooth was present on one or the other side of the midline, it would be an indication that the contralateral tooth has either been lost or did not develop. It is important to exclude other diseases before coming to the conclusion that the patient definitely has SMMCI. Any condition where two maxillary central incisor teeth began normal
Sonic Hedgehog (SHH) gene at 7q36 may be associated with SMMCI, but recent studies suggested that the existence of several other candidate genes including loss of function affecting the mouse Gas1 gene is associated with microform HPE.[7] An extensive literature review of gene mutations in SMMCI patients, however, showed that most were in the HPE genes: SHH, SIX3, ZIC2, TGIF, GLI2, and PTCH.[7]

### Conclusion

The following main traits were found in the reported individual: indistinct philtrum, arch-shaped upper lip, absence of frenulum of the upper lip, mid-palatal ridge, and nasal obstruction or deviation. These findings help us to arrive at the diagnosis of SMMCI. Furthermore, craniofacial profile[2] including neurocranium[9] and craniofacial[9] morphology should be incorporated in the criteria applied in diagnosing an SMMCI patient. While diagnosis of the presence of SMMCI is easy, it is difficult to understand the pathogenesis behind this type of agenesis, although it is obvious that a genetic component is involved along with environmental factors. The present pace of genetic research might unveil the cause and pathogenesis of SMMCI syndrome in the near future. Management of a patient with SMMCI is interdisciplinary, including neuropsychiatric diagnosis,[2,10] otolaryngologists, and plastic surgeons when nasal obstructive anomalies are found.[8] If true short stature is suspected, the patient should be referred to a pediatric endocrinologist and the need of growth hormone administration can be determined.[8] A pediatric dentist, an orthodontist, and a prosthodontist might constitute delivering of good dental care throughout life. A pediatric consultation and a referral to a geneticist are crucial for further investigation, as was done in this reported case.

### Acknowledgments

We extend our special thanks to the following doctors: Dr Hitesh Desai (MD Pediatrics), Dr Rajesh Korde (MD Pathology), and Dr Shailaja Saxena (Medical Geneticist).

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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