CASE REPORT

Solitary Median Maxillary Central Incisor Syndrome: A Case Report

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Abstract

Aim: This report intends to present a case of solitary median maxillary central incisor syndrome (SMMCI) and its multidisciplinary team approach for diagnosis of other associated anomalies, with special emphasis on their management.

Background: Solitary median maxillary central incisor syndrome is a unique developmental condition characterized by only maxillary central incisor and a series of developmental defects, appearing as a syndrome. The appearance of a single incisor may take place due to the union of two incisor teeth or the absence of tooth germs. The mechanism of the fusion is still uncertain.

Case description: A 9-year-old female child reported with a chief complaint of pain in the right lower back tooth for the past 10 days. The presence of a single maxillary central incisor was an incidental finding. Then a detailed history and multidisciplinary evaluations revealed the diagnosis of SMMCI syndrome.

Conclusion: The effort in diagnosing and managing this syndrome had a strong impact on the child's life of which the parent was highly motivated and got a better understanding of associated problems of overall development.

Clinical significance: In SMMCI syndrome, the patient requires a multidisciplinary health team in order to improve their quality of life. It is of greater importance to diagnose the syndrome and treatment of these median line deformities.

Keywords: Case report, Incisor, Solitary median maxillary central incisor syndrome, Syndrome.

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Background

The presence of only a central incisor is an unusual dental deformity, while the inherited absence of an upper central incisor is also an uncommon event, which was first reported by Scott in 1958.[1] It can occur either as a sequestered dental finding or it could be associated with a series of developmental defects, appearing as a syndrome. Hall et al., in 1997, had originally termed this syndrome as solitary median maxillary central incisor syndrome or SMMCI syndrome and reported a frequency rate of 1 in 50,000 live births with female gender predilection.[2] This is a unique developmental deficiency comprising median line structures of the head and body. The characteristic features of the syndromes are choanal atresia, median line stenosis or congenital pyriform aperture stenosis (PAS), cranial bones, maxilla, and its confined dentition mainly the central incisor tooth germ, and sometimes the brain (holoprosencephaly).[3]

The significant finding is the presence of a single central incisor in both primary and permanent dentitions, which is positioned exactly in the midline of the maxillary alveolus. The other characteristic features of this syndrome are an arch-shaped form of the upper lip with an inarticulate philtrum, high labial position, palatal suture malformation, abrupt palatal arch with projecting midpalatal ridge, and absence of labial frenulum and incisive papillae.[3–5] To improve the well-being of the patient, it is better they are treated with a multidisciplinary healthcare team as they require continuing comprehensive care.

The objective of this article is to report a case of an incidental outcome on a single maxillary central incisor leading to a multidisciplinary team approach for diagnosis of an SMMCI syndrome with specific emphasis on their management.

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Case Description

A 9-year-old female child born to phenotypically healthy parents reported to the department of pediatric dentistry with a complaint of pain in the right lower back tooth for the past 10 days; the pain was spontaneous in nature and nocturnal in type. Medical history shows that the child was diagnosed with a congenital cardiac problem soon after birth and underwent surgical repair for the same. No other medical history was reported. Dental history revealed that the child had undergone extraction of deciduous teeth a year back under physical restraints. There was no previous history of dental trauma. On general examination, the child was thin built with below average height (122 cm) and weight (25 kg), along with a dysmorphic face with relatively decreased intercanthal distance and an upturned nasal tip with a long philtrum. On close

observation and interaction, the child exhibited a slight alteration in their behavior, which was not appropriate for her age. On intraoral examination, there was a bifid upper labial frenulum, "V" shaped constricted maxillary arch, high arched palate, and numerous decayed teeth on both upper and lower arch, and some of the primary teeth exhibited preshedding mobility (Fig. 1). In addition to this, a unique clinical finding of a single central incisor positioned exactly at the center of the maxillary arch was noted (Fig. 1A). An intraoral occlusal radiograph and a panoramic radiograph were advised to assess the multiple carious teeth and single central incisor. The intraoral maxillary anterior occlusal radiograph and a panoramic radiograph confirmed the clinical findings of single maxillary central incisors (Fig. 2). In addition to this, it was noted that the maxillary permanent canines were highly placed near the nasal floor, raising a query whether that could be impacted, for which an orthodontic opinion was sought, cone-beam computed tomography (CBCT) was advised for better assessment of the position of canines. CBCT revealed that one central incisor was present, and canines had a favorable outcome for eruption (Fig. 3). Hence, on correlating the obtained information, we gave the provisional diagnosis of SMMCI.

A detailed history revealed the child had one maxillary central incisor in primary dentition, which was confirmed by the photographs of the child. As it could be associated with a spectrum of findings appearing as a syndrome, to confirm the associated findings, the child was referred for a multidisciplinary evaluation and opinion. After the evaluations and investigations by the multidisciplinary team, it was found that some systemic defects were associated with it (Table 1).

Correlating the clinical findings and radiographic investigations, a final diagnosis of SMMCI syndrome was provided. After obtaining concerned opinions and fitness, the treatment plan was framed. Since the patient lacked cooperative ability for multiple surgical extractions, we opted for full mouth rehabilitation under general anesthesia (Fig. 4).

Discussion

Solitary median maxillary central incisor syndrome is a phenotypically distinctive developmental anomaly of an indefinite event occurring during the 35th to 38th day *in utero*. The upper central incisor is characterized by its unique form and position. It occurs either as an isolated dental abnormality or concomitant with multiple median line defects.[6] The etiology of this syndrome remains uncertain, although some studies explain the chromosomal abnormalities, for example, deletion in chromosome 18 (18p), 47XXX, and *SHH* gene.[3] Some midline variances, such as CHARGE association[7,8] and the VACTERL association[2,9] were also found as a part of SMMCI. It was also found in ectodermal dysplasia,[10] Duane retraction syndrome,[11] oromandibular limb hypogenesis syndrome type I, hemifacial microsomia, and Goldenhar's syndrome.[12]

Congenital narrowing of the back of the nasal cavity, PAS, and intranasal stenosis can be associated with the dental findings of SMMCI. The nasal deformity in these syndromes is caused due to failure of the oronasal fragmentation, resulting in the congenital hindrance of the hard or membranous posterior nasal aperture. These abnormalities can be identified in prenatal diagnosis through magnetic resonance imaging (MRI) and complete diagnosis with computed tomography. Thus, numerous studies have observed the association between SMMCI and nasal obstructions. Lo et al. found congenital stenosis with SMMCI in 63% of patients.[13] Hall et al. observed a positive association with nasal congenital obstruction in 21 SMMCI patients.[3]

Chromosomal abnormalities were observed in SMMCI patients. Aughton et al.[14] and Dolan et al.[15] observed in SMMCI patients there was an omission of parts of 18p associated with mutation of the *SHH* gene. The occurrence of single central incisor could be due to the union of two adjacent teeth or agenesis of a tooth bud. The mechanism of the fusion is still uncertain, but

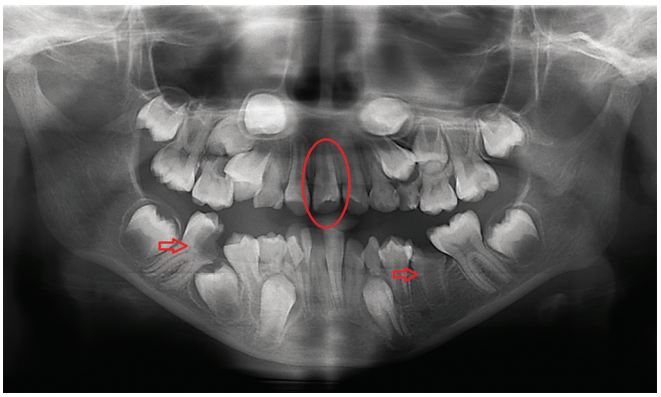
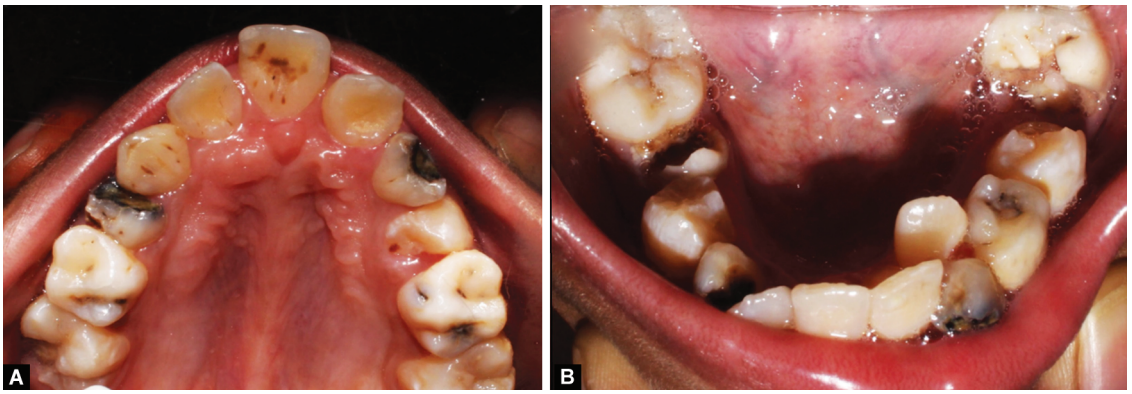


Fig. 2: Radiographic presentation at diagnosis of SMMCI: panoramic radiograph shows the presence of a single permanent maxillary central incisor exactly in the midline and multiple decayed teeth



Figs 1A and B: Clinical picture at diagnosis of a dental finding of SMMCI: (A) Maxillary occlusal view showing single central incisor positioned exactly at the midline of the maxillary arch; (B) Mandibular occlusal view showing multiple decayed teeth

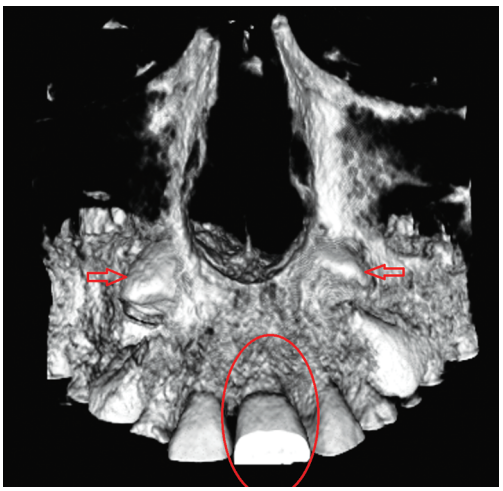
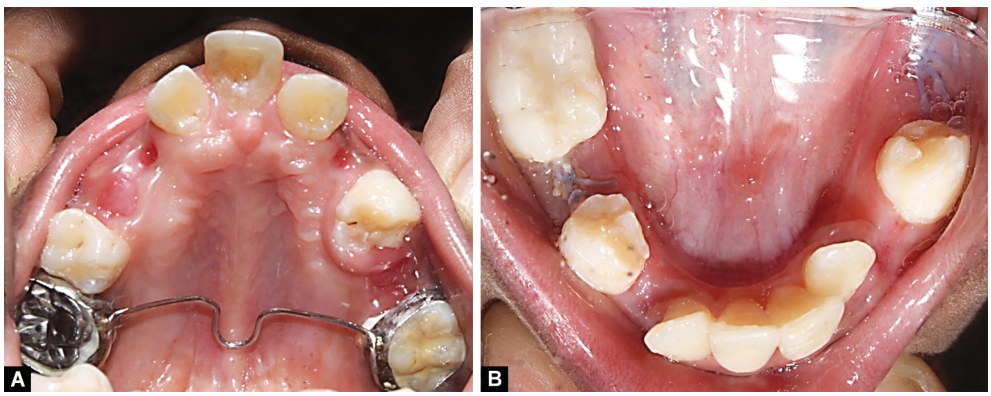


Fig. 3: Cone-beam computed tomography analysis for the maxillary canine's position and SMMCI; CBCT shows a favorable outcome for canine eruption and reconfirms the position of a single maxillary central incisor

Table 1: Consolidated multidisciplinary evaluation and findings

Sl. no.	*Multidisciplinary evaluation*	*Findings*
1.	Pediatrician	Growth—at risk (height 122 cm/below 3rd percentile; weight 25kg)
2.	ENT/otolaryngological evaluation	Submucosal cleft with bifid uvula, choanal atresia
3.	Endocrinological evaluation	Normal findings
4.	Psychological evaluation	Mental age—5 years with mild intellectual deficit
5.	Ophthalmological evaluation	Hypotelorism (mild variant)
6.	Neurological evaluation	MRI—partial agenesis of corpus callosum



Figs 4A and B: Postoperative clinical pictures after full mouth rehabilitation: (A) Maxillary occlusal view; (B) Mandibular occlusal view

it was believed that the critical nonexistence or reduction in lateral growth from the median line lead to the untimely union of the epithelial lamina.[16] As mentioned, SMMCI can occur as a sequestered finding, or it could be connected with a series of median line defects in the body involving autosomal dominant holoprosencephaly and growth retardation.[17] Becktor et al. assessed the maxillary growth in 11 patients with SMMCI and found that the anterior portion to incisive foramen on the median intermaxillary suture was affected, but the maxilla had normal horizontal and vertical growth.[18,19] Kjaer et al. evaluated the craniofacial morphology of SMMCI patients and observed there is an alteration in the sella turcica, small anterior cranial base, a short retrognathic, and posteriorly inclined mandible were evident in five patients. And these patients had characteristics, such as complete or incomplete midpalatal ridge, deviated nasal septum with nasal obstruction, and lack of frenum in the upper lip. This unblocks us that generally, SMMCI is associated with many median line and craniofacial deformities.

Early diagnosis of such condition is possible that is even before birth during routine midtrimester ultrasound, soon after the birth by a neonatologist by checking the head circumference, eyes, nose, or by a pediatric dentist at 8 months during the eruption of a primary central incisor or between 1 and 9 years of age on a routine dental examination.[17,20]

As soon as the diagnosis is made, it is mandatory that the child should be tested by a multidisciplinary healthcare crew along with long-term comprehensive care. It is also important to attend to the patient being it any kind; the patient has to be approached holistically. If there is any doubt in clinical situations, elect for further investigations for early detection with strong evidence-based decisions, and whenever necessary, it has to be managed with a multidisciplinary approach with a long-lasting follow-up for a healthier life.

Clinical Significance

This case report has a phenomenal clinical significance under the perception of early diagnosis and the importance of the role of healthcare teams like pediatric dentists, physicians, pediatricians, speech therapists, psychologists, and geneticists in the management of SMMCI syndrome patients.

Conclusion

The multidisciplinary team effort in diagnosing the syndrome resulted in parents' understanding of the child's associated developmental median line deformities. More importantly, after the psychological evaluation, they were suggested to change the school to a special school for better development of the child.

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