Case Report

Solitary median maxillary central incisor syndrome: A rare entity

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Abstract

Solitary median maxillary central incisor (SMMCI) syndrome is a complex disorder and a rare dental anomaly, which is estimated to occur in approximately 1:50,000 live births. It is a unique developmental abnormality which involves the central incisor tooth germs, occurring with or without systemic involvement; hence, its early diagnosis is of great importance. The objective of this study was to discuss a case of SMMCI syndrome and various dental treatment alternatives available in management of such cases. Due to the possible association of this syndrome with other developmental problems, an early diagnosis and management becomes important. Moreover, when it is associated with other developmental malformation and defects, the management becomes multidisciplinary.

Keywords: Dental anomaly, solitary median maxillary central incisor, syndrome

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INTRODUCTION

Solitary median maxillary central incisor (SMMCI) syndrome is a rare developmental disorder affecting the maxillary central incisor tooth germs. SMMCI has an occurrence rate of 1:50,000 live births with a higher incidence in women. SMMCI is characterized by a symmetrical single maxillary central incisor tooth located exactly in the midline of the maxillary alveolus.[1-3] The presence of solitary symmetrical central incisor was first reported by Scott.[4] Rappaport *et al.*[5] found a frequent association of short stature and solitary maxillary incisor and named the condition "monosuperoincisivodontic dwarfism." Since then, multiple cases of SMMCI with or without systemic involvement have been reported in the literature.

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Hall *et al.*[1] (1997) originally named this syndrome as "Solitary median maxillary central incisor, short stature, choanal atresia/midnasal stenosis syndrome", it is now shortened to "Solitary median maxillary central incisor syndrome" or SMMCI syndrome.[2] The etiology of SMMCI is believed to be unknown events occurring between the 35th and 38th days *in utero* involving midline structures of the head including the cranial bones, the maxilla, its dentition specifically central incisor, the nasal airways and sometimes the brain (holoprosencephaly [HPE]) with other midline structures of the body. SMMCI is seen in both deciduous and permanent dentition and is characterized by the presence of symmetric maxillary central incisor, the absence of labial frenum and incisive papilla with narrow nose. The palate is V-shaped with unusual narrow ridge along the midpalatal suture, extending

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from the incisive papilla region to the posterior border of the hard palate.[1-6] Several syndromes have been recorded in patients with SMMCI including CHARGE[7] and VACTERL associations, velocardiofacial syndrome,[8] autosomal dominant HPE,[9] ectodermal dysplasia,[10,11] Duane retraction syndrome,[12] Goldenhar syndrome,[13] and oromandibular limb hypogenesis syndrome type 1.[3]

There are only a few cases of isolated SMMCI reported in the literature. The present case report describes a rare case of isolated SMMCI syndrome and various dental treatment alternatives for management of such cases.

CASE REPORT

A 9-year-old female patient reported to the orthodontic clinic with a chief complaint of unesthetic appearance due to the presence of a single large upper front tooth. Medical and dental history revealed that she was born full term and had no history of any tooth loss due to caries or trauma. Extraorally, the patient had dolichocephalic, symmetric face with harmonious facial thirds [Figure 1]. The central portion of the upper lip was high with an indistinct philtrum which is characteristic of SMMCI syndrome. On profile examination, the patient had a convex profile with a decreased nasolabial angle and a retrognathic chin.

On intraoral examination, the patient was in a mixed dentition stage with one large maxillary central incisor situated exactly in the midline [Figure 2]. Maxillary and mandibular arches were symmetrical with angles Class I molar relationship. The patient had a V-shaped palate and marked midpalatal vomerine ridge. The labial frenulum of the upper lip and incisive papilla were absent. A periapical radiographic examination showed a central incisor having only single root canal [Figure 3].

Clinical and radiographic examination both confirmed that the child had a symmetrical permanent solitary maxillary central incisor. No caries lesions or restorations were present. The nasal cavity was narrow with slight asymmetric nasal septum which is characteristic of SMMCI syndrome. In terms of other syndrome-associated anomalies, the patient had no intellectual disability or any other developmental anomalies.

Treatment alternatives

As the main concern of the patient was esthetics, the only way to achieve this in patients with SMMCI is to provide space for the second central incisor. At appropriate age, space can be created by an orthodontist using expansion appliance to widen the palate, thus providing sufficient space for the SMMCI tooth to be moved to one side of midline and creating space for contralateral missing central incisor to be replaced with prosthesis. After creation of space, the incisor may be replaced using fixed prosthodontic

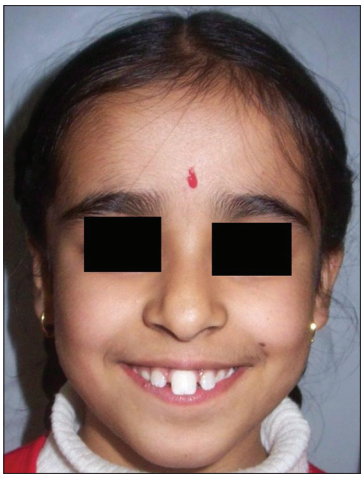

Figure 1: Extraoral frontal view showing facial appearance of the patient with solitary median maxillary central incisor syndrome

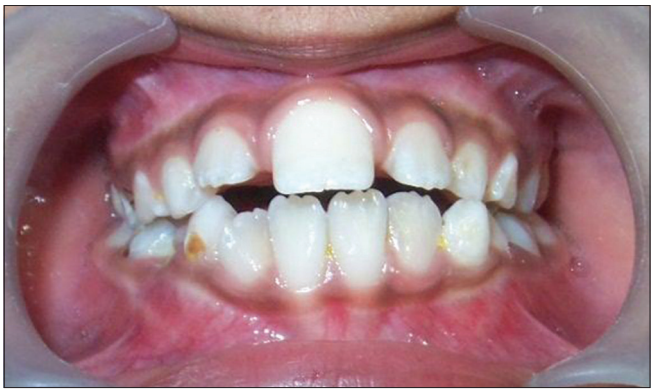

Figure 2: Intraoral frontal view of anterior teeth showing solitary symmetric midline permanent central incisor tooth and absence of labial frenum

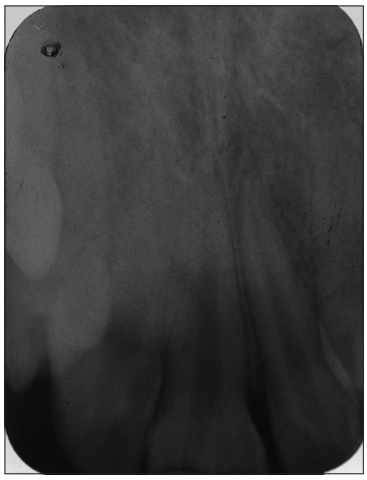

Figure 3: Intraoral periapical radiograph revealing solitary maxillary central incisor with one root and root canal

rehabilitation. Among the fixed prosthodontic treatment, options are single-tooth implant, Maryland retainers and bridge. Fixed partial dentures/bridge are not indicated in adolescents. A single-tooth implant is also not indicated unless alveolar growth is complete as it may result in implant position which is different from adjacent normal teeth which keep on growing in adolescents. Maryland retainers with minimal tooth preparation can be used in such cases but are often associated with fracture and failure. A modified removable orthodontic retainer with artificial central incisor is the easiest treatment option available till the time growth is complete and an implant or bridge can be used. After completion of growth (17–18 years of age), SMMCI tooth can be recontoured using a labial veneer to create the anatomical form of appropriate size and shape along with prosthodontic rehabilitation of missing teeth.

DISCUSSION

The SMMCI tooth is a rare developmental disorder characterized by unique form and position of the maxillary central incisor. Hall *et al*.[1,2] speculated that a critical absence or deficiency of lateral growth from the midline –a deficiency due to lack of normal cell division at the critical time for midline structures, on or about gestation day 37 or 38, results in premature fusion of the spreading dental lamina from the left and right sides in the maxilla, thus preventing the formation of two complete and separate central incisor teeth. Instead, one tooth consisting of two normal distal halves of the central incisors develops from inductive epithelium and mesenchymal condensations of these two tooth germs.

Till date, the exact pathogenic or genetic defect of SMMCI is undetermined. However, there is some evidence, showing that isolated SMMCI can result from mutation in the sonic hedgehog gene on the long arm of chromosome 7.[14] Deletion of parts of chromosome 18 (18p) in patients with SMMCI was reported by Dolan *et al*.[15] and Aughton *et al*.,[16] and deletion in the terminal portion of chromosome 7q was reported by Masuno *et al*.[17]

SMMCI can occur as an isolated anomaly or in combination with other morphologic defects, such as hypotelorism, indistinct philtrum, absence of the frenulum of the upper lip, vomerine ridge and nasal obstruction or septal deviation.[18] The involvement of SMMCI was first reported by Scott who described a girl with the presence of a solitary median maxillary central incisor as an isolated finding. Fulstow[6] reported a case of SMMCI, wherein apart from the single central incisor, the patient was having short stature, congenital heart disease, microcephaly and scoliosis. There have been studies in the literature that found no correlation between SMMCI and systemic changes. Wesley *et al*.[19] reported two cases of SMMCI in patients with normal stature, and Cho and Drummond[20] reported three cases of SMMCI with no systemic involvement and growth deficiencies. Clinical significance of SMMCI lies in its association with HPE, which is a lethal disorder when fully expressed. As SMMCI itself has been thought to be the mildest form of HPE, the offspring of patients with isolated SMMCI are at risk for HPE.[21]

SMMCI could be associated with various congenital nasal cavity anomalies such as choanal atresia, midnasal stenosis and nasal pyriform aperture stenosis.[1,2] Alis and Ward[21] evaluated six patients with congenital stenosis of the nasal pyriform aperture and found that four of these had SMMCI.[2] Lo *et al*.[22] found that 63% of the patients with congenital stenosis in the nasal pyriform aperture also presented SMMCI, whereas Hall *et al*. found that all the 21 patients with SMMCI had a history of nasal congenital obstruction, 7 had choanal atresia and 8 had intranasal stenosis.[1]

SMMCI can be associated with deviations in neurocranial size and shape and in craniofacial morphology. The neurocranial and craniofacial morphology of 13 children with SMMCI was evaluated by Tabatabaie *et al*.[23] using profile radiographs and cephalometric analysis. They observed that the size of the neurocranium, the maxillary prognathia, the maxillary inclination, the mandibular prognathia and the inclination of the mandibular incisors are significantly reduced in SMMCI.

Till date, very few cases of SMMCI as an isolated anomaly have been published in the literature which may be because of the reason that patients with SMMCI and systemic abnormalities have been reported by both medical and dental professionals whereas isolated SMMCI is reported mainly by dentists. Early diagnosis of SMMCI is important because it may be a sign of other more severe congenital abnormalities. Successful dental treatment of patients with SMMCI requires comprehensive treatment planning and a multidisciplinary team of pediatric dentist, prosthodontist and orthodontist.

CONCLUSION

A case of SMMCI should alert the clinician toward the possible associations. A complete and thorough evaluation of any patient with SMMCI should be done, and the earliest possible diagnosis and management of the dental and other co-existing developmental anomalies should be done.

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.

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Conflicts of interest

There are no conflicts of interest.

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