Solitary median maxillary central incisor in association with hemifacial microsomia: A rare case report and review of literature

ASHOK UTRJEA, SYED NAVID ZAHID, RICHA GUPTA

Abstract
Solitary median maxillary central incisor (SMMCI) is a rare dental anomaly. It is estimated to occur in 1:50,000 live births. The SMMCI tooth differs from the normal central incisor in that the crown form is symmetric and it develops and erupts precisely in the midline of the maxillary dental arch in both primary and permanent dentitions. Presence of SMMCI with hemifacial microsomia (HFM) is a very rare clinical condition. We report a case of HFM in a male of Indian origin who presented with SMMCI in both primary and permanent dentitions. The association of HFM with SMMCI may be due to defective development of neural crest cells and/or lack of space in maxilla.

Keywords: Hemifacial microsomia, solitary median maxillary central incisor, anodontia

Introduction
Solitary median maxillary central incisor (SMMCI) is a rare dental anomaly. Presence of a solitary symmetrical maxillary central incisor of normal crown dimensions situated precisely in the midline in both primary and permanent dentitions was apparently first reported by Scott. Since then, SMMCI has been reported both as an apparently isolated dental finding and with a variety of midline developmental defects, e.g. holoprosencephaly (HPE), and/or pituitary dysfunction. The name SMMCI was originally suggested by Hall et al. Various terminologies like “monosuperoincisivodontic dwarfism,” “single central incisor syndrome,” “single maxillary central incisor,” and “single incisor,” suggested by other authors, do not adequately describe the peculiarly formed incisor tooth. The SMMCI differs from normal incisor in that the crown form is symmetric and it develops precisely in midline of maxillary dental arch in both primary and permanent dentitions and is not a supernumerary tooth. The incidence of SMMCI is around 1:50,000 live births. SMMCI may occur in isolation or in association with other systemic abnormalities like short stature, pituitary insufficiency, microcephaly, choanal atresia, midnasal stenosis, and congenital nasal pyriform aperture stenosis. It is considered as one of the most minimal expressions (microforms) of the HPE spectrum. Deletions on chromosomes 7 and 18 (at 7q36.1 and 18p-), which are in chromosomal regions that harbor HPE genes, have been reported to be associated with SMMCI. Some investigators found Sonic Hedgehog (SHH) mutation in patients affected by SMMCI. Hehr et al. emphasized the wide phenotypic variability in families with HPE and SHH mutation.

Hemifacial microsomia (HFM) is believed to be the second most common craniofacial anomaly following cleft lip and palate, involving first and second branchial arch derivatives. An association has been described between HFM and hypodontia. Barring a single case report, the presence of single maxillary central incisor has not been reported in association with HFM. This article reports the case of a boy with HFM syndrome who presented with the classical signs of the syndrome and an SMMCI.

Case Report
A 6-year-old boy of Indian origin attended cleft and craniofacial clinic at Oral Health Science Centre, PGIMER, Chandigarh, with the chief complaint of asymmetry of face. The baby was born to non-consanguineous healthy parents of normal stature. There was no history of hereditary disease in his family. There was no history of medication during pregnancy and the delivery was atraumatic and full term.

The patient had undergone surgical procedure 1 year after birth for torticollis and had average built and normal mental development. There was no history of nasal malformation or stenosis.

The face was asymmetric with hypoplastic left side. Microtia was apparent with preauricular tags. The philtrum was...
indistinct and occlusal plane was canted up to left. The masseter and temporalis muscles were hypoplastic and mastoid prominence was small [Figure 1].

Intraoral examination revealed the presence of single deciduous central incisor. The incisor was placed in midline and had symmetrical right and left contours, which resembled distal contour of central incisors. There was no labial frenum and incisive papilla. The palate had characteristic v shape, with fine bony ridge running along the length. The patient had poor oral hygiene and multiple carious teeth. The arches were narrow in size with the left side more collapsed than the right [Figure 2].

Intraoral periapical x-ray views of the patient’s maxillary incisor region confirmed the presence of solitary incisor. The permanent incisor was seen above the deciduous and erupting in midline. The intermaxillary suture was distinctly visible on the radiograph [Figure 3].

The patient required a multidisciplinary management and long-term follow-up. In the first phase, oral hygiene was monitored and oral hygiene instructions reinforced. This was followed by restoration of all carious lesions by a pediatric dentist.

The patient was reviewed every 6 months and radiographs for regular monitoring of facial growth were done. The facial asymmetry aggravated as the age advanced due to differential growth of right and left sides [Figures 4–6]. At the age of 8½ years, the permanent maxillary central incisor which has the morphology of fused central incisors erupted in midline. The size of this central incisor was comparable to normal central incisor [Figure 7]. Panoramic view showed that the coronoid process, condylar process, and mandibular ramus were hypoplastic on the left side and eruption of teeth was delayed due to lack of space in arch [Figure 8].

As a part of dental treatment, maxillary expansion can be commenced in late mixed dentition and the space created after moving SMMCI to one side of the arch can be replaced by a single tooth implant or crown by a prosthodontist. Mandibular distraction and reconstruction of face can be planned in multiple stages. Consultation with ENT specialist for auricular defects should be done. Genetic counseling is required, as some individuals with SMMCI, normal intelligence, and normal brain image have had children with HPE and SMMCI has been recognized as a risk factor for holoprosencephalic offspring.

Discussion

To the best of our knowledge, this is the second case report in literature in which SMMCI was found in association with HFM. The first was reported by Garcia de Paula e Silva et al. in a 10-year-old male patient. Hall et al. reported a series of 21 consecutive cases of SMMCI, the
SMMCI can also be a feature of recognized syndromes or associations or a finding in patients with specific chromosomal abnormalities. Nanni et al.[9] reviewed the extensive number of anomalies in addition to HPE with which SMMCI had been reported. The correlation has been found with, Velocardiofacial (VCF), DiGeorge syndrome, HPE, ectodermal dysplasia, and Duane retraction syndrome, but not with HFM.

HFM is a syndrome which affects craniofacial structures to varying degree. This syndrome occurs at a higher rate in males than in females.[20] The incidence of this condition, also known as oculoauriculovertebral dysplasia, is about 1 in 5000 to 25,000 live births.[21] The phenotype is highly variable and the features include unilateral deformity of the external ear and small ipsilateral half of the face with epibulbar dermoid and vertebral anomalies. Coloboma of the upper eyelid is frequent. The ear deformities range from preauricular tags of cartilaginous masses to atresia of the external auditory canal, anomalies in the size and shape of the external auricle, and even anotia.[22] In addition to the craniofacial anomalies, there may be cardiac, vertebral, and central nervous system defects.[16] Commonly observed malformations include microtia, macrostomia, and failure of formation of mandibular ramus and condyle. The patient had classical features of HFM.
A factor aggravating asymmetry at an earlier age might be torticollis, but due to early release (within 1 year), the effect on growth of face is expected to be minimum.

The etiology of both the conditions remains uncertain although various theories have been proposed. The SMMC may be due to a congenitally missing tooth bud with agenesis of the incisor and the remaining incisor erupts in the midline. It has been hypothesized that the formation of one instead of two teeth could result from a disturbance in the mitotic potential of the incisor tooth bud, which could be under genetic and environmental determinants. It has also been proposed that for SMMC to form, the dental lamina must have fused prematurely in midline, resulting in apposition and fusion of forming tooth buds. This prevents normal formation of intervening bone and associated soft tissue. It has also been suggested that space limitation in maxillary arch or deficiency of lateral growth from midline would result in premature fusion of spreading lamina from the right and left sides. In this patient, the solitary incisor might be a result of premature fusion in midline as in HFM there is lack of space in jaws.

Although genetic and environmental pathogenic mechanisms have been proposed for HFM, it is likely that the cause is multifactorial. In a review of various pathogenic models, the authors underscore studies of the genetic component of HFM. Several investigators favor a vascular basis for the development of this anomaly. Another theory is based on disruption of migration of neural crest cells during craniofacial development. If a disturbance in neural crest cell development or migration plays a role in HFM and hypodontia, it would follow that a correlation between the two conditions should occur. It is generally accepted that normal odontogenesis requires the presence and interaction of neural crest ectoderm and mesenchymal cells. Disturbances in the odontogenic process can produce abnormal or incomplete dental development. In a previous study, an association has been found between smaller size of dentition and HFM.

It should be noted that the SMMC may occur alone or in other conditions bearing no relationship to HPE. Cohen states that rather than thinking about a single central incisor as a microform of HPE, it is better to think of it as

1. An integral component of severe HPE,
2. An anomaly that occurs alone and in other conditions unrelated to HPE,
3. The only manifestation in some members of a dominantly affected family with variable expressivity of HPE and incomplete penetrance, and
4. Rarely as an isolated dominant trait with an SHH mutation.

In this case, SMMC along with HFM may be considered as an anomaly that has occurred alone and is unrelated to HPE. It is also possible that in patients presenting with HFM, the SMMC may be more than a chance finding. A further critical evaluation of patients with HFM and association between the two might be helpful in providing insight into the unknown etiology of the two conditions.

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