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Case Series

Rare cases of (SMMCI) solitary median maxillary central incisor syndrome: Identification and management

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ABSTRACT

Condition where there is agenesis of central maxillary incisor is unique and very rare with association of peculiar stomatognathic features is called (SMMCI) Solitary median maxillary central incisor syndrome which may or may not have systemic involvement, and the condition is found to affect 1:50,000 live births. SMMCI is a very rare abnormality in developing stage of fetus involving the tooth germs of central incisor. The most common systemic association is with holoprosencephaly, and diagnosis in early stage is of utmost importance for managing dental aesthetic issue patient generally suffers from the syndrome. The objective of current article is to discuss two cases of SMMCI syndrome; one with systemic involvement having esophageal Artesia, a rare entity with SMMCI syndrome and the other one without systemic involvement. In both cases, patient's parents were not aware of the dental anomaly the child was suffering, and parents were highlighted on treatment options in managing such dental cases. As there are a significant number of SMMCI syndrome cases with systemic involvement and other developmental problems, it becomes prudent to have a quick and prompt diagnosis in initial stages of life and simultaneously the management approach becomes multidisciplinary.

1. Introduction

Solitary median maxillary central incisor syndrome (SMMCIS) is a unique and infrequent anomaly of stomatognathic system, with a 1:50,000 live births occurrence rate and higher incidence in females, aborted fetuses and still births. SMMCIS was initially described by Hall et al. in 1997. These cases are generally referred to a dental surgeon for aesthetic reasons.[1,2] Patients suffering from this syndrome have chances of being carriers of HPE (Holoprosencephaly), and patients having SMMCI without any systemic involvement and thereby making the condition a potential risk factor for HPE. HPE is a malformation of congenital origin, features includes impaired cleavage of the embryonic forebrain in association with varying degrees of facial dysmorphism.[3]

The Etiological factor of SMMCIS is believed to be the unknown in-utero events occurring between 35th and 38th week, with main involvement of midline structures of the head and body including the skull bones, the maxilla sometimes mandible its dentition specifically the central incisor, the nasal cavity and airways and sometimes the brain (holoprosencephaly) [HPE] and sometimes Gastrointestinal tract. Both deciduous and permanent

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dentitions are involved in SMMIC and are characterized by the symmetric presence of a single maxillary central incisor, absent labial frenum and incisive papilla with slight nose deformity. V shaped palate along with a narrow ridge of midpalatal suture, covering the whole midpalatal suture of the hard palate.[4–6]

As SMMCIS is a rare condition and there is a paucity of cases reported in the literature. More insights are required into the condition as it may or may not be associated with systemic involvement. The present article described two cases of SMMCIS with and without systemic involvement its identification, diagnosis and various dental treatment alternatives for management of the condition.

2. Case Report 1

A 4 year old female patient reported to the Dental Department with a chief complaint of carious teeth and wanted to get them restored. Medical and dental history was not significant. Extra orally, the patient had a symmetric face with no distinct facial deformities. High upper lip placement with an indistinct philtrum was present making the striking features of SMMCI syndrome. The patient had a convex profile and a decreased nasolabial angle.

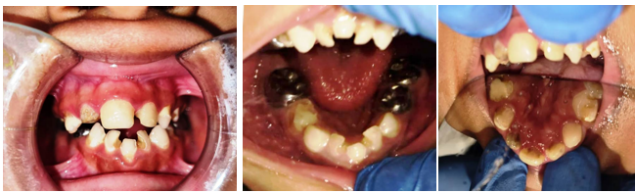

Figure 1: Solitary deciduous maxillary and mandibular central incisor situated in the midline.

On intraoral examination, the patient was in a primary dentition stage with solitary deciduous maxillary and mandibular central incisor situated exactly in the midline [Figure 1] and multiple carious teeth. Maxillary and mandibular arches were narrow and symmetrical with an end-on molar relationship. Incisive papilla as well as the labial frenum was absent. Intraoral periapical radiographic examination showed a deciduous central incisor having only a single root canal and the presence of a permanent single central incisor tooth bud [Figure 2]. The nasal cavity was narrow with a slight asymmetric nasal septum which is characteristic of SMMCI syndrome without systemic involvement. Finally, after taking parents' consent, carious teeth were restored and pulpectomy was performed in tooth no. 55, 75, 84 and 85 [Figure 3]. Oral hygiene instructions were given but patient did not come for regular follow-ups and restorations of remaining mutilated teeth. In terms of other syndrome associated anomalies, the patient had no intellectual disability or any other developmental anomalies. Patient guardians were informed about the condition as they were not aware of the same and future treatment

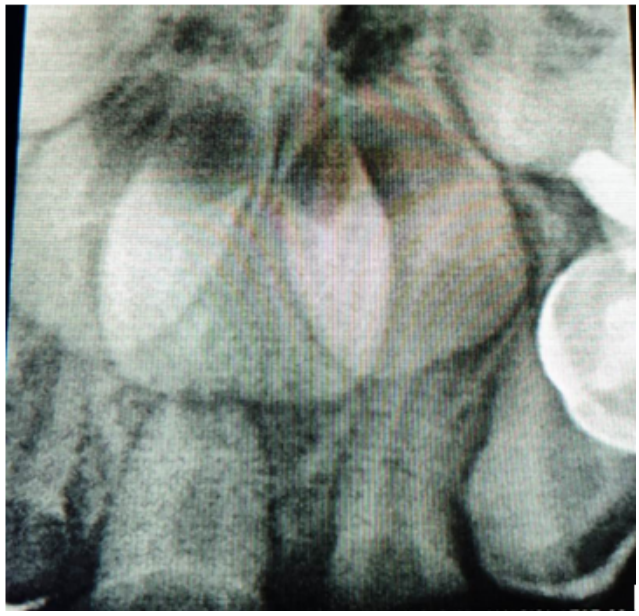

Figure 2: IOPA showing permanent single central incisor tooth bud.

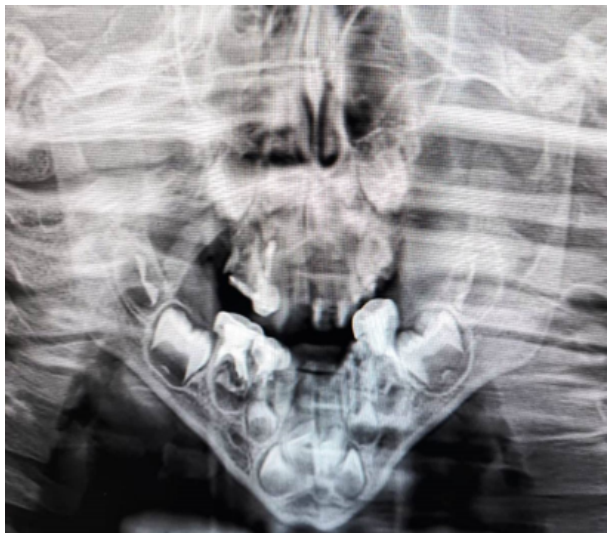

Figure 3: OPG showing treated (pulpectomies) 55, 75, 84 and 85.

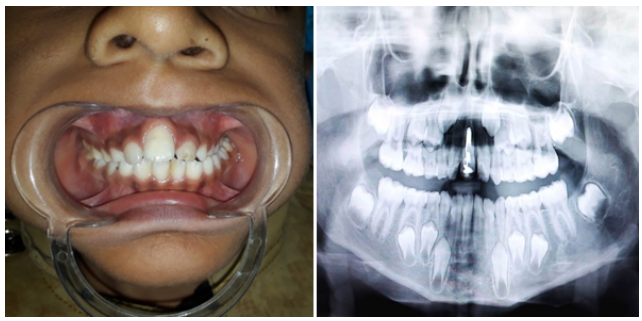

Figure 4: Intraoral and OPG showing symmetrical permanent solitary maxillary central incisor.

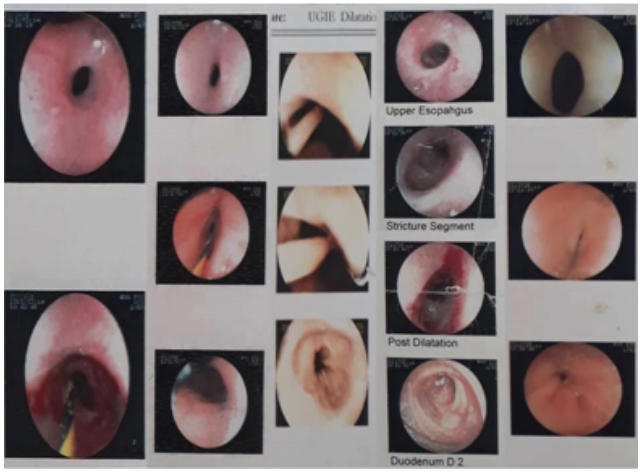

Figure 5: Upper gastrointestinal endoscopy dilatation procedure.

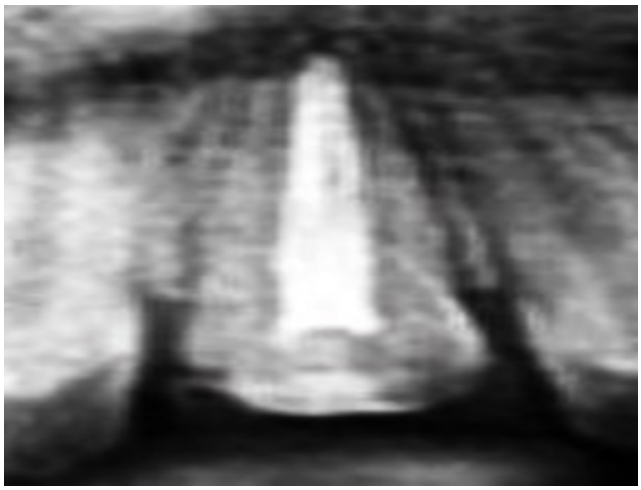

Figure 6: Endodontically treated solitary maxillary incisor.

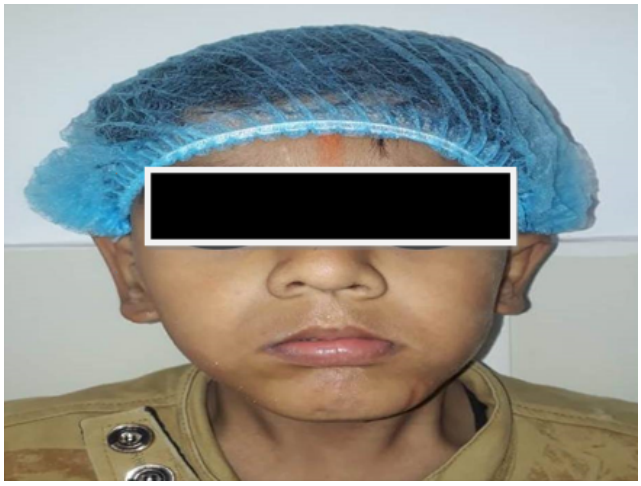

Figure 7: Extra oral picture showing indistinct philtrum

alternatives regarding age appropriate esthetics treatments with the involvement of a pediatric dentist, orthodontist and prosthodontist at a specific age of child.

3. Case Report 2

A moderately built 10-year-old boy born from healthy parents was referred to the Pedodontic department for evaluation of pain due to tooth trauma in the upper front tooth region. The patient had history of a fall 1 month back and has been complaining of pain since then. Treatment started in a private clinic, as pain was not subsiding; the patient was referred to us. On examining clinically and on conducting radiographic evaluation, it was found that the child had a symmetrical permanent solitary maxillary central incisor with tenderness on percussion and grade 1 mobility with no fracture of root as well as coronal region (Figure 4). The final diagnosis of Ellis class V fracture was made and the patient was advised to undergo endodontic treatment of the tooth. Parents were introduced to the dental anomaly the child was facing and on taking a detailed history of the patient it was found that the patient had difficulty in feeding with respiratory trouble after birth and from medical records of the patient it was found that the patient had upper esophageal stricture at 10-12 cm of esophagus and was operated post natally followed by 14 cycles of UGIE (Upper Gastrointestinal Endoscopy) dilatation procedure for esophageal stricture over a period of 2 years (Figure 5). The patient was having no difficulty with eating in his present state and had no respiratory and cognitive disabilities and difficulty. Finally, after taking parents' consent, endodontic treatment was completed over a period of week and pain was relieved and the tooth was stabilized (Figure 6). Skeletal Class I relation and a unilateral crossbite resulting from a narrow maxilla. Additionally, an indistinct philtrum, a prominent midpalatal ridge and the absence of labial frenulum were evident (Figure 7).

4. Discussion

Scott in 1958 was the 1^{st} clinician to describe SMMCIS as an isolated finding,[5,6] followed by Fulstow in 1968. In 1976, "Monosuperocenroincisivodontic dwarfism" term was introduced by Rapport et al. SMMCI syndrome was accepted as a term finally to describe the condition where symmetric placement of a single maxillary central incisor tooth is present with and without other systemic involvement.[7] Most of the time both primary and secondary dentition is involved. For precise diagnosis of patients with SMMCI syndrome, some unique identification features should be fulfilled. Solitary tooth must exist precisely on the mid-palatine raphae. It is important to exclude other conditions which may give features of the presence of a single central incisor before coming to the conclusion

that the patient definitely has SMMCI which may include conditions like tooth lost due to trauma, periodontal problem or caries. SMMCI may be presented as an isolated finding or in association with many other midline developmental anomalies. SMMCI patients may be involved with more than 70 systemic anomalies or without any recognized syndrome. Conditions may be congenital heart disease, short stature, congenital nasal pyriform aperture stenosis, pituitary insufficiency, mid ;nasal stenosis, cleft lip and/or palate, less frequently, microcephaly, Gastrointestinal involvement, hypopituitarism, and hypothyroidis, with short stature in almost 50% of the cases.[8] Various syndromes have been found to be associated with patients having SMMCI condition including velocardiofacial syndrome, autosomal dominant HPE, ectodermal dysplasia, triple X syndrome, and Duane retraction syndrome. Moreover, recently it was found that, SMMCI syndrome is also association with oromandi bular; limb hypogenesis syndrome type I in the literature.[7]

Nanni et al. suggested that a missense mutation in the Sonic Hedgehog (SHH) gene at 7q36 may be associated with SMMCI although the etiology of SMMCI condition is not precisely certain. Gene mutation studies conducted in SMMCI patients, showed that most patients were in the HPE genes: SHH, SIX3, ZIC2, TGIF, GLI2, and PTCH.[8] Mutation in the SHH homolog gene may be associated with SMMCI, however latest studies suggested that the existence of several other target genes including loss of function affecting the mouse Gas1 gene is associated with microform HPE.[9] SMMCI syndrome may occur to developmental disturbance occurring during gestation (35th-38th days approximately) in the maxilla, with abnormal formation of tooth germs and alveolar bone.[10]

5. Conclusion

The following main common traits were found in both individuals: indistinct philtrum, arch; shaped upper lip, absence of frenum of the upper lip, mid palatal ridge, and nasal deviation with systemic involvement in one case. All these finding were conclusive of SMMCI syndrome diagnosis, inclusion of the craniofacial profiling like neurocranium and craniofacial morphology can be included into the criteria applied to diagnose an SMMCI patient. Although to diagnose SMMCI is easy, its pathogenesis is quite ambiguous with obvious genetic and environmental factors involvement.

A multidisciplinary effort is required to manage patients with SMMCI syndrome; timely diagnosis helps in management of patients with suitable outcomes.

The stomatognathic treatment plan for both cases described consisted an initial phase including rehabilitation of carious and endodontically involved teeth, and finally an esthetic phase that would require combination of orthodontic treatment and prosthodontic rehabilitation.

6. Declaration of Patient Consent

All appropriate patients' consent forms were taken before writing the article, which includes patient(s) written consent from the legal guardian of the patients for his/her/their images, medical records and other clinical information. Patients and their legal guardians understood that all precautions were taken for data integrity and identification of patients.

7. Source of Funding

None.

8. Conflict of Interest

None.

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