Concomitant solitary median maxillary central incisor and fused right mandibular incisor in primary dentition

G. Shilpa, Sivakumar Nuvvula, Niraj Gokhale, V. Yamini

Abstract

Solitary median maxillary central incisor (SMMCI) is a unique developmental anomaly in primary dentition. It involves central incisor tooth germs and may or may not be associated with other anomalies. Its presence, concomitant with fusion of right mandibular incisors has not previously been reported. A 5-year-old girl was presented with a single symmetrical primary maxillary incisor at the midline, with the absence of labial frenulum, an indistinct philtrum and a prominent midpalatal ridge. There was an associated fused tooth in the right incisor region and radiographic examination confirmed only one maxillary central incisor in both the dentitions. Family history revealed that the father of the girl also had a similar anomaly providing probable evidence of etiological role for heredity in SMMCI.

Keywords: Central incisor, double tooth, maxillary, median, solitary median maxillary central incisor

Introduction

Solitary median maxillary incisor (SMMCI) is a rare anomaly with a reported prevalence of 1 in 50,000 live births and is more common among females.[1] The most common clinical presentation in the oral cavity is the presence of a single central incisor at the midline of the maxilla in both dentitions. The etiology of SMMCI is unknown, but it may be related to a disruption in the development of the maxilla, which occurs at approximately 35-38 days of intrauterine life, with abnormal formation of tooth germs and alveolar bone or soft tissue.[2-4] It is known to be associated with anomalies of midline structures of brain and face, growth retardation (with or without growth hormone deficiency), malformations of sella, choanal atresia, mid-nasal stenosis, and pyriform aperture stenosis.[1,5,6] The anomaly occurs due to disruption in development of maxilla leading to lack of space and premature fusion of dental lamina.[3,4] Cases of isolated SMMCI without any associated anomalies have also been reported.[7] It could also form part of a syndrome well known as Solitary median maxillary incisor syndrome (SMMCIS) which includes symmetrical central incisor, absence of labial frenulum and incisive papilla, absence of inter-maxillary suture in front of the incisive fossa, and a prominent mid-palatal ridge.[8] It may be associated with holoprocencephaly,[9,10] Goldenhar's syndrome,[11] and oromandibular-limb hypogenesis syndrome type 1.[12] We report a case of SMMCI in a 5-year-old girl concomitant with fusion of right mandibular incisors.

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Case Report

A 5-year-old girl, only child, born to nonconsanguineous parents was brought to our out patient clinic with complaint of a large anterior tooth in the maxillary arch. On clinical examination the child was of normal built (weight: 20 kg, height: 112 cm, and head circumference: 49 cm). The philtrum was indistinct. Intraoral examination revealed an absent maxillary median labial frenulum with a large, symmetrical, midline tooth, and fused right mandibular incisors [Figure 1]. The incisive fossa was prominent along with a very prominent midpalatal ridge [Figure 2]. The intraoral periapical radiograph revealed only one central incisor with single root and root canal and no impacted tooth but with only one developing succedaneous tooth [Figure 3a]. Intraoral periapical radiograph of mandibular incisors confirmed fusion of right primary central and lateral incisors [Figure 3b]. Family history revealed that her father also had a similar anomaly with midline single maxillary incisor.

Discussion

The prevalence of double tooth in various studies ranges from 0.1% to 4.1%. The prevalence rate shows a definite geographical variation, the Chinese and Japanese having high prevalence rates.[13] It is more common in the anterior mandibular region of primary dentition. Most frequent combination in double tooth includes central and lateral incisor or lateral incisor

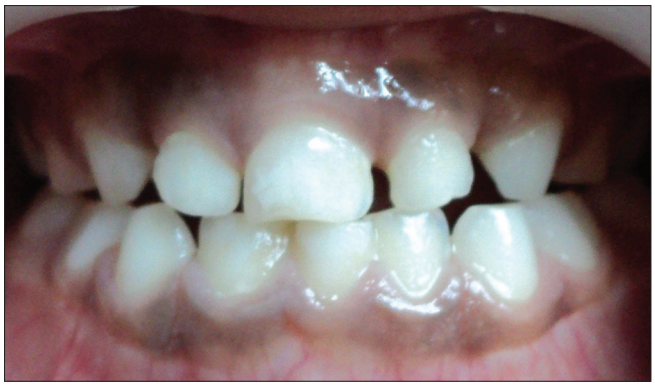

Figure 1: Clinical picture showing large median maxillary incisor with absent labial frenulum and fused mandibular right incisor

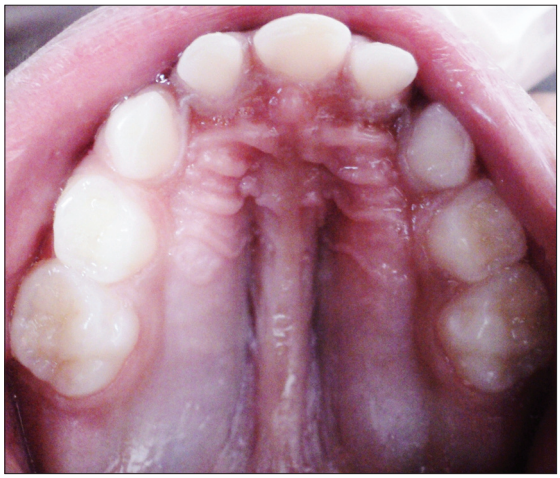

Figure 2: Intraoral picture shows prominent midline palatal ridge

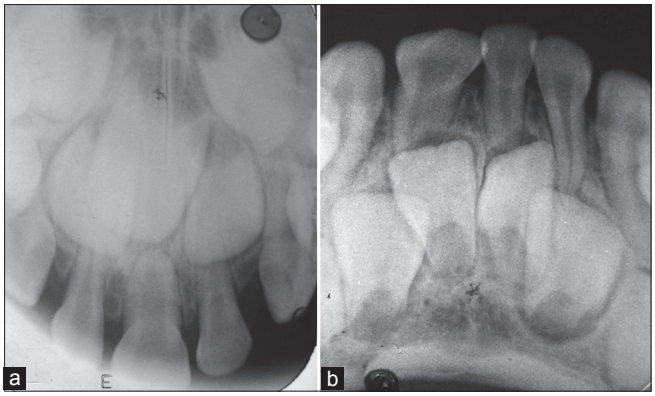

Figure 3: Intraoral periapical radiographs revealing solitary central incisor with one root and root canal and one succedaneous tooth (a), and fused mandibular right primary incisors (b)

and canine. Double tooth involving maxillary central incisors in primary dentition is very rare. It can present as a solitary finding SMMCI, solitary median maxillary central incisor syndrome (SMMCIS) or it can be a mild manifestation along with multisystemic major anomalies. Traumatic loss, fusion of primary or permanent central incisors with a supernumerary tooth and mesiodens must be ruled out before diagnosing SMMCI. In conditions where two maxillary central incisors began normal development but failed to proceed beyond the cellular developmental stage cannot be diagnosed as SMMCI.[7] The case presented by us showed double tooth (fusion) in the mandibular right incisor region.

Hitchin and Morris[14] have shown that persistence of interdental lamina (which maintains continuity between tooth germs) initiates formation of double tooth. Several cases of double teeth in families have been reported implicating a hereditary cause. Various etiological factors have been implicated for SMMCI. Disruption of maxilla during intrauterine period of 35–38 days may lead to lack of space and premature fusion of dental lamina across the midline resulting in SMMCI.[5,6] Solitary median incisor is seen in all patients with holoprosencephaly which is a midline developmental defect of prosencephalon and face.[10] Mutations in human sonic hedgehog gene can cause holoprosencephaly.[15] It has an autosomal dominant inheritance with 70% penetrance.[16] Solitary median incisor could also be associated with CHARGE (coloboma, heart defects, choanal atresia, retarded growth and development, genital anomalies, and ear anomaly), VACTERL (vertebral anomalies, cardiac anomalies, trachea-oesophageal fistula, nasal and labial anomalies), Goldenhar's syndrome, hypothalamic hamartoma, triple-X syndrome, ectodermal dysplasia, hypotelorism, and mid-axial defects of craniofacial and brain structures.[6,7,11]

Children with SMMCIS could have growth and mental retardation. There are cases of SMMCIS with short stature and growth hormone deficiency. However, Cho and Drummond in their report of three cases of SMMCIS have reported normal growth.[17] Stanhope *et al*,[18] felt that growth abnormalities could manifest at a later stage. Hall *et al*.[1] suggest regular follow up of SMMCIS children associated with at least 2 SD below the mean height for their age and gender by an endocrinologist. They have demonstrated abnormal sella turcica in their cases. Pituitary function and sellar structure may or may not be associated; in the case reported by Bolan *et al*., sella turcica was normal.[2] Though the SMMCIS is shown to be associated with learning difficulty there have been various reports with normal intellectual development.[2,4]

SMMCIS is sometimes associated with midline nasal cavity abnormalities like choanal atresia, nasal stenosis, and aperture stenosis. These children may or may not manifest obstruction symptoms. In the case reported by Velasco *et al*. there was severe asphyxia at birth.[19] Hall *et al*. and Kjaer *et al*.[1,10] have found that clinically symptomatic obstruction is present in only few of their patients with neonatal nasal obstruction. Hall *et al*.[1] recommend evaluation by an otolaryngologist once a diagnosis of SMMCIS is made. Bolan *et al*.[2] have reported "open trachea" causing breathing difficulty in their case.

Chromosomal deletions have been shown in chromosomes no. 7, 18 and 22 in some cases of SMMCIS.[1,15] In the two cases of SMMCIS reported by Yassin and Bolan[2,20] in one of the twins, the other twin was normal. In the online database developed by John Hopkins University this SMMCIS is referred as Online Mendelian Inheritence in Man[21] (OMIM 147250). Our report involving the child patient and her father with manifestation in both of them supports possible X-linked dominant inheritance apart from autosomal inheritance pattern, though this needs further proof of genetic analysis for which the family did not give consent.

SMMCIS can be associated with posterior cross bite,[1,4,22] Beckor *et al*,[23] have dislocated the mandible on the right side because of occlusion repositioning provoked by contact with a canine tooth. Some authors recommend treatment with orthodontic appliances at the permanent dentition stage to gain space for implant-prosthodontic rehabilitation.[6,7,24]

Johnson *et al*.[25] used magnetic resonance imaging (MRI) for prenatal diagnosis of solitary median maxillary central incisor syndrome and reported that MRI provided detailed insight into fetal anatomy and pathology, aiding prenatal diagnosis, and facilitating parental counseling.

Conclusion

SMMCIs should alert the clinician towards possible associations. These children require evaluation by endocrinologist and otolaryngologist. Genetic studies are required to establish the possible mutations. Parents should be counseled regarding the need for regular follow up by the multidisciplinary team.

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