Solitary Median Maxillary Central Incisor Versus Agenesis of the Maxillary Central Incisor

Fabrício Kitazono de Carvalho, DDS, MS, PhD [1]
Juliana Arid, DDS, MS [2]
Andiara De Rossi, DDS, MS, PhD [3]
Francisco W.G. Paula-Silva, DDS, MS, PhD [4]
Paulo Nelson Filho, DDS, MS, PhD [5]

ABSTRACT

A solitary median maxillary central incisor (SMMCI) is rare and affected individuals may carry a potentially serious condition known as SMMCI syndrome. However, many of these cases do not receive proper attention because they are misdiagnosed as agenesis of the maxillary central incisor. The purpose of this manuscript is to report two cases of children with only one maxillary central incisor and draw diagnostic differences between the entities. A correct diagnosis is very important because if an SMMCI is confirmed, the patient should be referred for genetic counseling. (J Dent Child 2016;83(1):XX)

Received October 22, 2014; Last Revision November 25, 2014; Revision November 25, 2014.

KEYWORDS: SOLITARIAN MEDIAN MAXILLARY CENTRAL INCISOR (SMMCI), TOOTH AGENESIS, CENTRAL INCISOR

Solitary median maxillary central incisor (**SMMCI**) syndrome is a developmental disturbance that involves midline structures of the face and is presented in all cases by the occurrence of an SMMCI, probably due to mutations in the Sonic hedgehog gene (locus 7q36).[1-3] SMMCI presents peculiarities that differentiate this tooth from an ordinary central incisor, such as the fact that it emerges in the midline, has a symmetric crown, and occurs in both primary and permanent dentitions.[4] Patients who have an SMMCI present an arched upper lip, undefined or short philtrum, narrow palate, absent or abnormal intermaxillary sutures in the region of the incisor foramen, and absent upper labial frenulum and incisive papilla.[3,4] The occurrence is rare (approximately one in 50,000 live births).[5]

An SMMCI may appear as an isolated dental anomaly or as part of other developmental disorders of midline structures such as obstruction of the nasal cavity, orbital hypotelorism, cerebral malformation, and microcephaly.[4,6] It may also be part of the spectrum of holoprosencephaly, a developmental defect of midline structures of the face resulting from total or partial failure of the division of the frontal lobe of the brain.[4,6] Malformation ranges from mild to severe; miscarriage usually occurs in severe cases, and facial malformation and intellectual disability occur in mild cases.[2-4] SMMCI can also be associated with other syndromes, including VATER association (Vertebral defects, Anal atresia, Tracheoesophageal fistula with esophageal atresia, and Radial or Renal dysplasia), CHARGE syndrome (Coloboma, Heart

[1]Dr. Carvalho is an assistant professor, [2]Dr. Arid a graduate student, [3]Dr. Rossi is an associate professor, [4]Dr. Paula-Silva is a graduate student, and [5]Dr. Nelson Filho is a professor, all in the Department of Pediatric Dentistry, School of Dentistry, University of São Paulo, Ribeirão Preto, São Paulo, Brazil.
Correspond with Dr. Arid at juliana_arid@hotmail.com

Anomaly, Choanal Atresia, Retardation, Genital and Ear Anomalies), and Goldenhar syndrome, among others.[4-6]

The human body is completely interconnected, so abnormalities in the oral cavity may represent signs and symptoms of systemic diseases;[7,8] therefore, an early diagnosis of SMMCI is important because it alerts to the need of an assessment of other serious developmental anomalies. It may also indicate that affected individuals carry genetic changes that can cause serious manifestations in their descendants.[9] Dental professionals play an important role in the early diagnosis of SMMCI, although they frequently misdiagnose it as agenesis of the maxillary central incisor.[6,9,10]

Differential diagnosis should be done by eliminating the probable causes of agenesis of the central incisor, including endocrine disorders, trauma, local or systemic infection, and congenital or genetic factors.[10] Moreover, unlike the SMMCI, the central incisor present in cases of maxillary central incisor agenesis presents a common shape, with distinct mesial and distal thirds of the facial surface, and is located lateral to the midline (i.e., left or right.[11] It may also not occur in both dentitions. Additionally, the incisive papilla and upper labial frenulum are present.[10]

The purpose of this manuscript is to report two cases of children with only one maxillary central incisor and draw diagnostic differences between the entities. A correct diagnosis is very important because if an SMMCI is confirmed, the person should seek genetic counseling.

CASE REPORT

CASE 1: SMMCI. A 10-year-old boy with Goldenhar syndrome presented to the Clinic for Special Care Patients at the School of Dentistry of Ribeirão Preto, University of São Paulo, Ribeirão Preto, Brazil, for dental care with a complaint of absence of a maxillary incisor. The patient was born to healthy nonconsanguineous parents after an uneventful full-term pregnancy. His mother denied using drugs during pregnancy. His height was within normal limits for his age. There was no history of orofacial trauma.

Goldenhar syndrome, also known as hemifacial microsomia or oculoauriculovertebral dysplasia commonly presents ocular, auricular, and vertebral malformations due to poor development of the first and second branchial arches.[6]

The extraoral examination revealed facial asymmetry and an indistinct philtrum. The intraoral examination revealed a solitary median maxillary central incisor (Figure 1a) and absence of upper lip frenulum and incisive papilla (Figure 1b). Clinically, the SMMCI presented indistinct mesial and distal surfaces. A Class II molar relationship was observed.

The panoramic radiograph confirmed the existence of an SMMCI in the midline, with incomplete root formation. Impacted teeth (maxillary left second premolar, mandibular right canine, mandibular first and second right premolars) and a hypoplastic left coronoid process of the mandible were also present. (Figure 2).

CASE 2: MAXILLARY CENTRAL INCISOR AGENESIS. A 12-year-old girl presented to the School of Dentistry at the University of the State of Amazonas, Manaus, Brazil, with a complaint of severe dental crowding. Her medical and dental were non-contributory. Her height was within normal limits for her age.

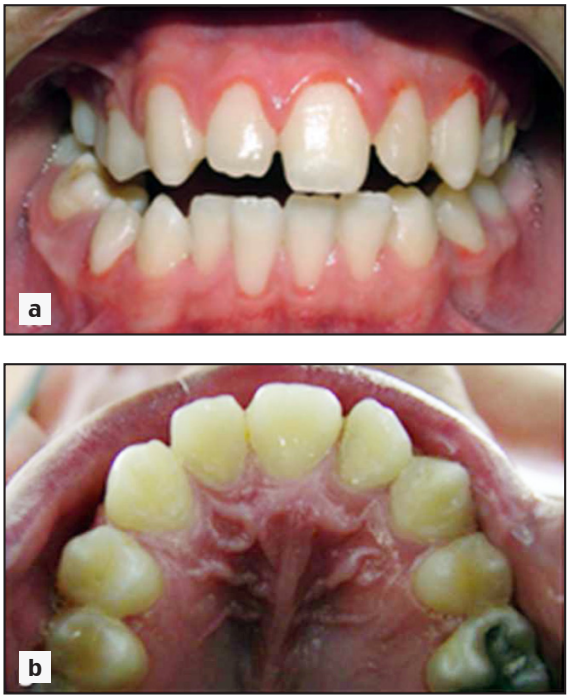

Figure 1. (a) Note a solitary median maxillary central incisor presenting indistinct mesial and distal faces and absence of upper labial frenulum. (b) Intraoral occlusal view showing absence of incisive papilla.

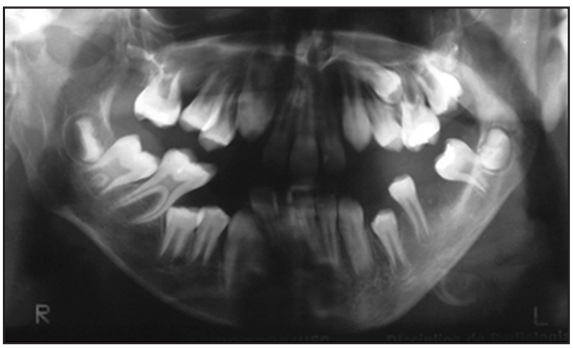

Figure 2. Panoramic radiograph confirming the existence of a solitary maxillary central incisor. No other dental anomalies were observed.

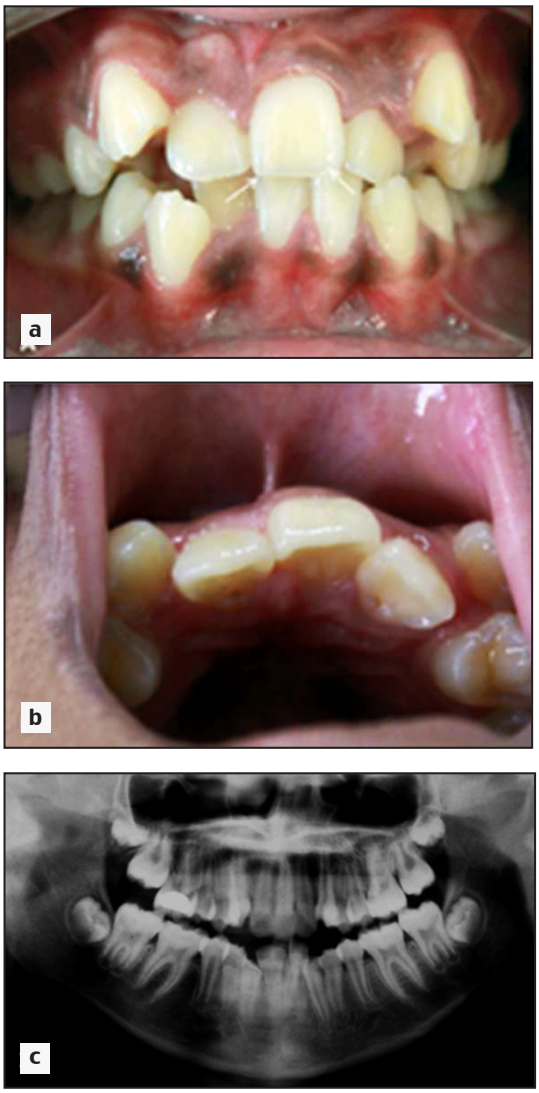

Figure 3. (a) Note only one maxillary central incisor with distinct mesial and distal faces. (b) Intraoral occlusal view showing the incisive papilla and maxillary labial frenulum. (c) Panoramic radiograph confirming the absence of the maxillary right central incisor with the maxillary left central incisor located laterally to the intermaxillary suture. No other dental anomalies were observed.

The clinical examination revealed only one maxillary central incisor (Figure 3a), with distinct mesial and distal surfaces, located laterally to the upper labial frenulum (Figure 3b) and presenting a shape similar to a normal maxillary left central incisor. The incisive papilla was present. A Class I molar relationship was observed; however, due to severe crowding, the patient was sent for orthodontic treatment.

The panoramic radiograph confirmed the absence of the maxillary right central incisor, with the maxillary left central incisor located laterally to the intermaxillary suture (Figure 3c). No other dental anomalies were detected.

DISCUSSION

This report presents two distinct cases of congenital absence of maxillary central incisors: an SMMCI and a solitary maxillary right central incisor due to the agenesis of the left counterpart. The patient with the SMMCI did not present a distinct philtrum, maxillary labial frenulum, or incisive papilla, which is in agreement with previous reports.[1-6] Furthermore, the SMMCI was symmetrical (i.e., the proximal portions of the facial surface were similar to the distal face of a normal maxillary central incisor). On the other hand, in the patient with agenesis, it was possible to distinguish the proximal distinct features of the facial surface of the central incisor. The fact that the tooth was not exactly in the midline, and the presence of the incisive papilla, philtrum, and the maxillary labial frenulum confirmed the diagnosis.[1,2,4,10]

Knowledge of the clinical features of these two entities is crucial because the presence of an SMMCI can be linked to several important systemic manifestations and can be passed on to the patient's offspring. Unfortunately, most dentists treat cases of SMMCI as agenesis.[1,6] These patients may present undiagnosed malformations and substantial mutations in their genome.

Because both conditions are found in the anterior region of the oral cavity leading to poor esthetics, the dentist is often the first professional sought by patients and should, therefore, be able to make the differential diagnosis between an SMMCI and agenesis of the maxillary central incisor.[4,12] In cases of diagnosis of an SMMCI, the dentist should provide the appropriate dental treatment and also refer the patient for a thorough medical evaluation, which should include neurological, cardiac, endocrine, and genetic assessment.[1-6,9]

REFERENCES

1. Blackmore K, Wynne DM. A case of solitary median maxillary central incisor (SMMCI) syndrome with bilateral pyriform aperture stenosis and choanal atresia. Int J Pediatr Otorhinolaryngol 2010; 74(8):967-9.
2. Viana Eda S, Kramer PF, Closs LQ, Scalco G. Solitary median maxillary central incisor syndrome and holoprosencephaly: a case report. Pediatr Dent 2010;32(5):424-7.
3. Lygidakis NN, Chatzidimitriou K, Petrou N, Lygidakis NA. Solitary median maxillary central incisor syndrome (SMMCI) with congenital nasal puriform aperture stenosis: literature review and case report with comprehensive dental treatment and 14 years follow-up. Eur Arch Paediatr Dent 2013;14(6):417-23.
4. Hall RK. Solitary median maxillary central incisor (SMMCI) syndrome. Orphanet J Rare Dis 2006;1:12.

5. Hall RK, Bankier A, Aldred MJ, Kan K, Lucas JO, Perks AG. Solitary median maxillary central incisor, short stature, choanal atresia/midnasal stenosis (SMMCI) syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1997;84(6):651-62.
6. Garcia de Paula e Silva FW, de Carvalho FK, Diaz-Serrano KV, de Freitas AC, Borsatto MC, de Queiroz AM. Solitary median maxillary central incisor in association with Goldenhar's syndrome: a case report. Spec Care Dentist 2007;27(3):105-7.
7. Carramolino-Cuellar E, Tomas I, Jimenez-Soriano Y. Relationship between the oral cavity and cardiovascular diseases and metabolic syndrome. Med Oral Patol Oral Cir Bucal 2014;19(3):e289-e294.
8. Islam NM, Bhattacharyya I, Cohen DM. Common oral manifestations of systemic disease. Otolaryngol Clin North Am 2011;44(1):161-82.
9. Marini M, Cusano R, De Biasio P, et al. Previously undescribed nonsense mutation in SHH caused autosomal dominant holoprosencephaly with wide intrafamilial variability. Am J Med Genet A 2003;117a(2):112-5.
10. Tichler HM, Abraham JE. Management of a congenitally missing maxillary central incisor. a case study. N Y State Dent J 2007;73(2):20-2.
11. Antal M. Replacing Congenitally missing central incisor: a case report with a special method to achieve the optimal emergence profile. Dentistry Case Reports. Surgery Curr Res 2013;S11:1-4. Available at: "http://dx.doi:10.4172/2161-1076.S11-003".
12. Ribeiro LA, Richieri-Costa A. Single median maxillary central incisor, hypophyseal tumor, and SHH mutation. Am J Med Genet A 2005;136A (4):346-7.