Cranio-facial manifestations of Solitary Median Maxillary Central Incisor Syndrome: case report

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Abstract

The median solitary maxillary central incisor syndrome (SMMCI) is a rare developmental disorder consisting of multiple defects found on the body midline. The aim of this report was to describe multiple craniofacial features of a 10-years-old girl presented with SMMCI, and discuss the dental treatment alternatives in such young patients.

Case Report

Cranio-facial manifestations of Solitary Median Maxillary Central Incisor Syndrome: case report

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SUMMARY

The median solitary maxillary central incisor syndrome (SMMCI) is a rare developmental disorder consisting of multiple defects found on the body midline.

The aim of this report was to describe multiple craniofacial features of a 10-years-old girl presented with SMMCI, and discuss the dental treatment alternatives in such young patients.
Keywords: SMMCI, children disease, Dental anomalies, Interceptive orthodontics

Introduction:

The present case report was prepared according to the CARE Guidelines.

Solitary median maxillary central incisor syndrome (SMMCI) is a rare developmental abnormality occurring in utero from the 35th-38th day after the conception.

The SMMCI is a very complex malformation involving different development defects of the midline structures. These development abnormalities occur in utero from the 35th-38th day after the conception. Defects may involve cranial bones, maxillary bone, airways (mid-nasal stenosis, choanal atresia, nasal pyriform) and midline brain structures associated with an increased risk of pituitary malformation and malfunction.

The etiology remains unknown although Nanni et al. suggested that this syndrome may be associated with a missense mutation in the SHH gene (I111F) at 7q36.

The presence of an SMMCI tooth can predict associated anomalies and in particular a serious anomaly which is holoprosencephaly.

Common congenital abnormalities may be also associated with SMMCI such as: severe to mild intellectual disability, congenital heart disease, cleft lip and/or palate, less frequently microcephaly, hypopituitarism, hypotelorism, convergent strabismus, oesophageal or duodenal atresia, cervical hemivertebrae, cervical dermoid, hypothyroidism, scoliosis, absent kidney, micropenis and ambiguous genitalia.

Diagnosis should be made by eight months of age, but can be made at birth and even prenatally at 18–22 weeks from the routine mid-trimester ultrasound scan.

Eliminating the conditions where only one incisor is present (traumatic loss of one central incisor, fusion of a primary or a permanent central incisor with a supernumerary tooth, mesiodens erupting in the midline), there are no other known conditions where this characteristic form of incisor tooth occurs.

Management of SMMCI syndrome patients depends upon the individual anomalies present. The SMMCI tooth itself is mainly an aesthetic problem, which is ideally managed by combined orthodontic, prosthodontic and oral surgical treatment; alternatively, it can be left untreated.

The present case report aimed to describe the cranio-facial manifestations of SMMCI syndrome in a 10-years-old girl and discuss the dental treatment alternatives in such young patients.

Case report:

A 10 years-old girl was referred to the department of Pediatric and Preventive Dentistry, Monastir Dental Faculty, Tunisia, regarding the enhancing appearance of her teeth.

The young child chief complaint was the unaesthetic appearance due to the presence of a single large upper front tooth.

The patient general condition was good but she showed a very low self-esteem because of her poor teeth appearance.

The interrogatory of her parent revealed that they had noted the condition around 3 years of age when a solitary median deciduous maxillary incisor had erupted but they did not give focus on that. The physical examination showed a normal stature.

The extra-oral examination showed specific facial features such as a long and narrow face, pinched nostrils and a slight facial asymmetry to her right side. An indistinct philtrum with an atypical arch-shaped outline of the upper cupid’s bow associated with labial open bite were evident (Figure 1a).

The lateral view exhibited a convex profile (Figure 1b).
The intraoral inspection revealed that the patient was in mixed dentition, with a totally symmetrical, large central incisor positioned precisely in the maxillary midline with the absence of the upper labial frenum (Figure 2 a, b, and c).

Bilaterally, the first molars were in full Class II occlusion with a bilateral crossbite, and a 4 mm overjet which was assessed between the large unique maxillary incisor and the mandibular incisors associated to a large open bite (Figure 2d and e).

The patient presented also a swallowing disorder.

The lower midline was mildly deviated to the left side by 1.5 mm.

The panoramic radiograph evidenced a solitary median maxillary central incisor exactly in the maxillary midline, with deviation of the nasal septum to the left side and an age-typical development of all other permanent teeth (Figure 3a).

The cephalometric analysis revealed a Class II skeletal pattern (SNA : 87°, SNB : 78°, ANB : 9°, AoBo : 5 mm), an increased vestibular inclination of the maxillary and mandibular incisors (I /F : 125°, IMPA : 102°, I/i : 106°) and a normal skeletal divergence (FMA : 27°) (Figure 2b).

For more investigation, the patient was referred to the pediatric and neurologic consultations.

The cone-beam computed tomography of the facial bone was indicated.

The examination was performed on a Toshiba-type 128-slice scanner using volume acquisition without IV injection of PDC with multiplanar reconstruction.

The CBCT revealed an appearance in favor of a single midline incisor syndrome and there were not any other health issues (Figure 4).

On overall assessment and as no history of dental trauma with avulsion of a central incisor was reported, and all typical extraoral and intraoral traits of the Solitary Median Maxillary Central Incisor (SMMCI) syndrome phenotype were present, the patient was finally diagnosed with SMMCI syndrome.

A treatment plan was developed which the main objective was to improve esthetics and restore masticatory function of the young patient.

First, the patient was advised to brush her teeth with normal fluoride toothpaste.

Second, a swallowing rehabilitation combined with a palatal expansion to correct the posterior crossbite was performed.

Finally, the young patient was referred to the Orthodontic Department to correct the skeletal Class II and to create a pleasing symmetrical smile either by extraction of the SMMCI with space closure or by space opening associated to a prosthodontic replacement for the a maxillary central incisor with either an implant borne crown or a resin bonded bridge.

Discussion:

SMMCI syndrome is a complex disorder characterized by neurodevelopment defects of the midline structures.

If the history of dental trauma was not reported, the absence of a permanent central incisor may be considered a rare form of hypodontia. However, the association of anatomical abnormalities in the middle line is used to alert the clinician to the possible existence of SMMCI syndrome which may require specialist care.

SMMCI was initially reported by Scott who described a girl with the presence of a single maxillary central incisor placed in the median position, in view of an isolated observation.

Later, another cases of SMMCI reported in addition to the single central incisor, short stature, congenital heart disease, microcephaly and scoliosis.
Although, in the present case, differently from what previously reported in literature, SMMCI may be associated with other systemic disorders such as autosomal dominant holoprosencephaly, growth retardation and midline developmental defects\(^4,7,8\).

The etiology is uncertain but some studies such as those conducted by Nanni et al. 2001 have shown, following DNA sequencing, mutations of the Sonic Hedgehog (SHH) gene both in SMMCI syndrome and in holoprosencephaly frameworks \(^9\).

The diagnosis of SMMCI can be made prenatally at the 18th-22nd week owing to the mid-trimester routine ultrasound\(^4\).

Increasingly, presumptive diagnoses are being made at birth with confidence when the multiple defects are present \(^5,6\).

Once the primary SMMCI tooth has erupted, diagnosis can later be confirmed by a pediatric dentist clinically and radiologically at 7–8 months of age\(^6\).

Kjaer et al. in 2001 examined the clinical features and craniofacial morphology of 10 patients with SMMCI aged between 8 and 17 years.\(^10\)

Also Bertolacini et al. study in 2009 consisting of 11 patients with SMMCI, who underwent radiological investigation.\(^11\)

The results of the two studies showed that the craniofacial morphology of the patients with SMMCI compared to the normal craniofacial parameters showed hypoplasia of the anterior cranial base, a hypoplastic and post-rotated maxilla, and a retrognathic and post-moved jaw.

In addition, these groups of patients had features such as: nasal obstruction, septal deviation, absence of frenulum of the upper lip and incomplete mid-palatal suture\(^10,11\).

In the present clinical case the patient showed the same signs of manifest SMMCI which confirmed our diagnoses.

In this complex disorder, management and treatment are complex and delicate. First of all, the evaluation of the psychological impact that this syndrome has on the patient and how the clinician can improve it, is important. Secondly, an interdisciplinary evaluation is fundamental for a correct management of the case. The treatment can be either by extraction of the SMMCI with space closure or by space opening associated to a prosthetic replacement for the a maxillary central incisor with either an implant borne crown or a resin bonded bridge. Thus, an early orthodontic evaluation for a proper planning of the treatment timing is encouraged \(^7\).

**Conclusion**:

Early diagnoses of SMMCI grant more appropriate clinical follow-ups and an appropriate treatment which could have a great advantage on the young patient quality of life.

Pediatric dentists should recognize this disorder at an early age since it is characterized by specific orofacial manifestations and should not consider it as a simple dental anomaly in order to establish a proper treatment for these patients.

References:

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**CONFLICT OF INTEREST**

None of the authors has declared any conflict of interest or financial disclosures.

**CONSENT**

Written informed consent was obtained from the legal guardian of the patient for publication of this case and accompanying images.

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