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Case Report

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Cranio Facial Manifestations of Solitary Median Maxillary Central Incisor Syndrome: A Case Report

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Abstract

The Median Solitary Maxillary Central Incisor Syndrome (SMMCI) is a rare developmental disease comporting several defects found on the body midline.

The aim of this report was to describe multiple cranio-facial characteristics of a 10-years-old girl diagnosed with SMMCI, and discuss the dental treatment options in such young patients.

Keywords: SMMCI, Children Disease, Dental Anomalies, Interceptive Orthodontics

1. Introduction

This case report was planned 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 [1,2].

The SMMCI is a very complex malformation involving different development distortions of the midline structures. These development abnormalities occur in utero at 4 to 5 weeks of pregnancy. Defects may involve maxillary and cranial bones, nasal cavity (mid-nasal stenosis, nasal pyriform, choanal atresia) and midline brain structures associated with an increased risk of pituity malformation and malfunction [3,4].

The etiology remains unknown although Nanni et al. suggested that this syndrome may be associated with a mutation in the SHH gene (I111F) at 7q36 [5].

The presence of an SMMCI tooth can anticipate associated abnormalities especially Holoprosencephaly.

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

Diagnosis can be established not only at eight months of age, but also at birth and even prenatally at 18 to 22 weeks from the usual mid-trimester ultrasound scan.

The SMMCI tooth should be ubiquitous in any case and may be detected radiologically prior to its eruption. After eliminating the conditions where only one incisor is present such as traumatic loss of one central incisor, fusion of a primary or/and 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 [6].

Management of SMMCI syndrome patients depends upon the individual anomalies present. The SMMCI tooth itself is principally an aesthetic problem, which is can be managed by combined orthodontic, prosthodontic and oral surgical treatment, or can remain untreated.

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

2. Case Report

A 10 years old girl was referred to the department of Pediatric and Preventive Dentistry, Dental Faculty of Monastir, 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 central incisor.

The general condition of the patient was good however; she showed a self-doubt because of her poor teeth appearance.

The interrogatory of her parent revealed that they had noted the situation 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 uncommon arch shaped outline of the upper Cupid's bow associated with labial open bite were evident (Figure 1a).
The lateral view revealed a convex profile (Figure 1b).

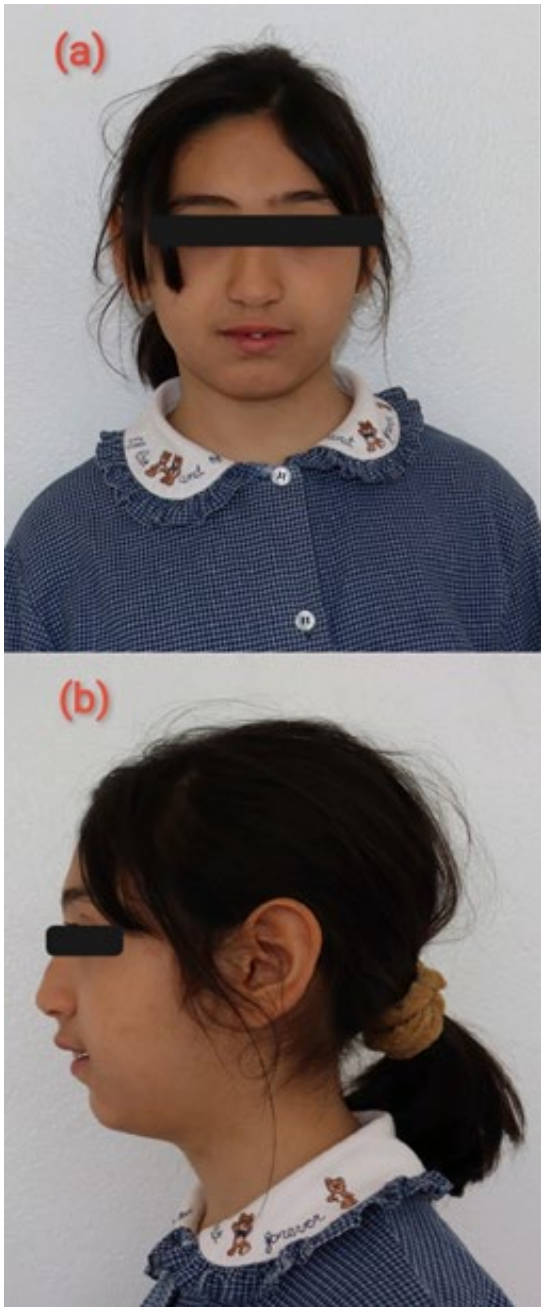

Figure 1: Extra oral examination (a) Front view (b) Profil view

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).

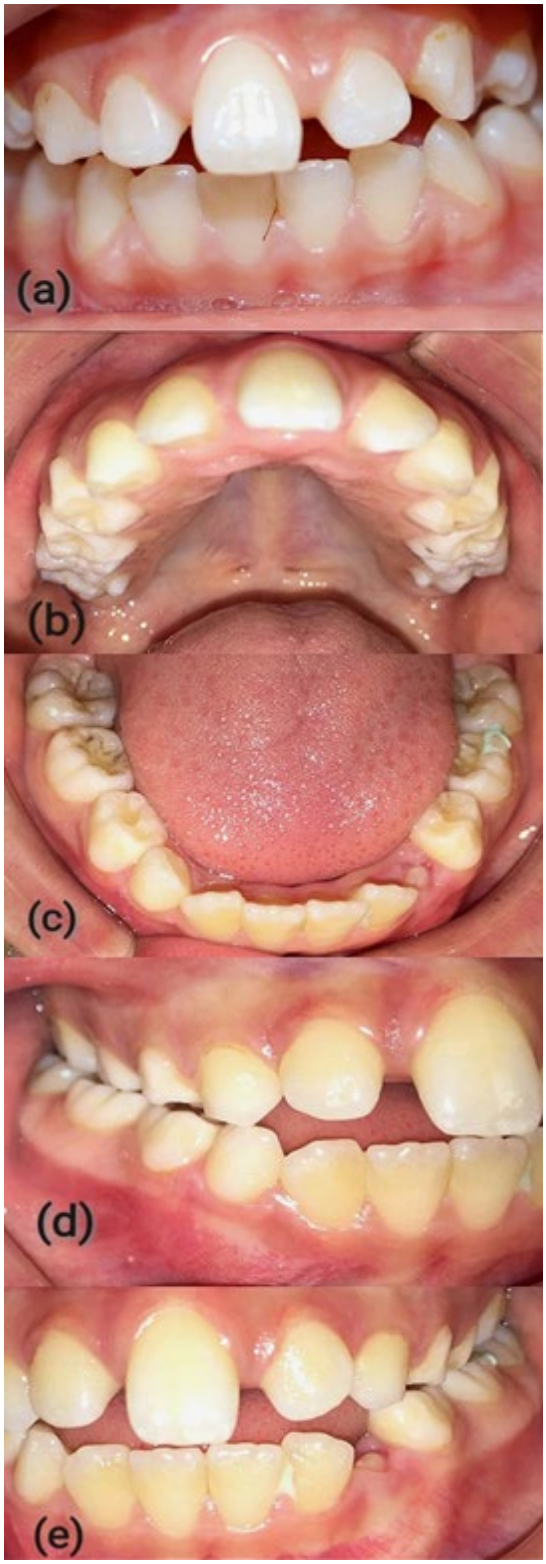

Figure 2: Intra oral examination (a) intra oral occlusal front view (b) intraoral maxillary view (c) Intraoral occlusal mandibular view (d) Right posterior occlusion view (e) Left posterior occlusion view

Bilaterally, the first permanent 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 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 3 a).

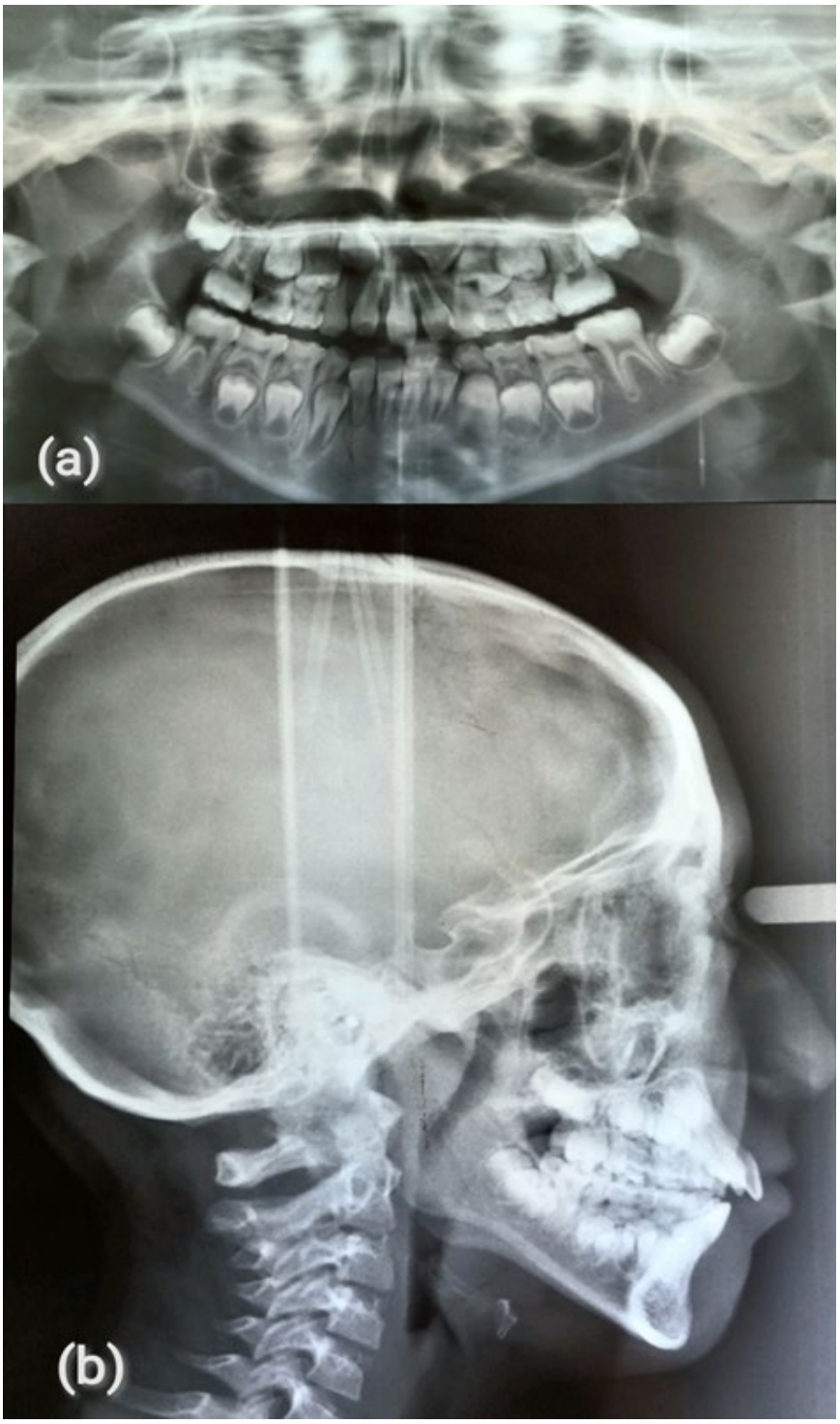

Figure 3: Radiographs. (a) Panoramic radiography (b) Lateral radiography

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

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).

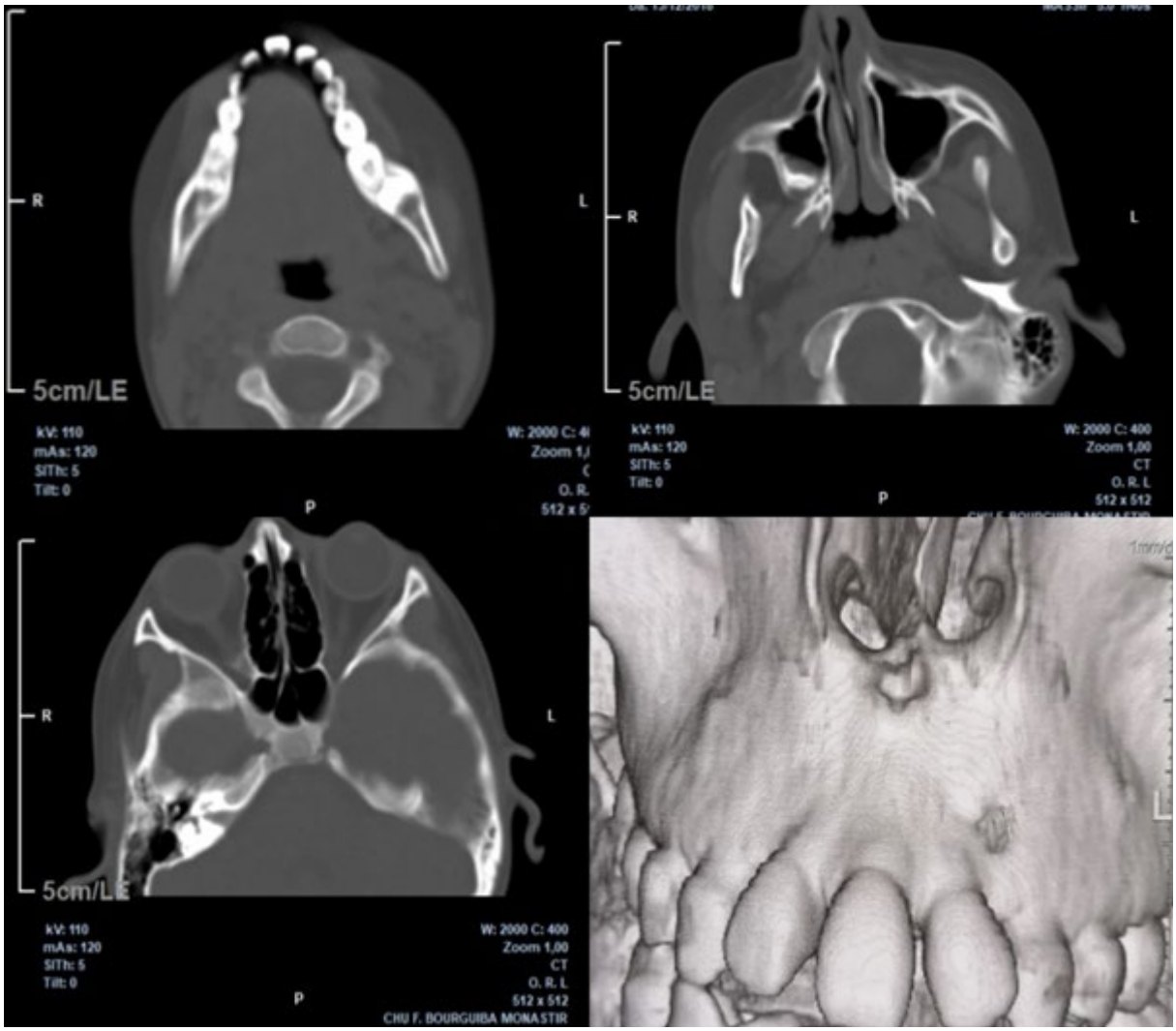

Figure 4: Craniofacial Computed tomography images

On overall assessment and as no history of dental trauma with avulsion of a central incisor was described, and all particular extra oral 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 purpose was to enhance aesthetics and restore masticatory function of the young girl.

First, the patient was encouraged 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 girl was addressed to the Orthodontic Department to correct the skeletal Class II and to create a pleasing symmetrical smile either by removal of the SMMCI with space closure or by space opening associated to a prosthodontic replacement for the

maxillary central incisor with either an implant borne crown or a resin bonded bridge.

3. Discussion

SMMCI syndrome is a complex disease characterized by neuro development defects of the midline structures6.

If no dental trauma was 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 described by Scott who described a patient with the presence of a unique maxillary central incisor placed in the midline, in view of an isolated observation [7].

Later, other cases of SMMCI reported in addition to the unique central incisor, short stature, microcephaly, scoliosis and congenital heart disease.

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 [8].

The etiology is unclear 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 to 22nd week owing to the mid trimester routine ultrasound.

More, presumptive diagnoses are being made at birth with confidence when the multiple defects are present.

Once the primary SMMCI tooth has erupted, a pediatric dentist can later confirm diagnosis clinically and radiologically at 7- 8 months of age.

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 underwengt radiological investigation [11].

The outcomes 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 retro gnathic 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.

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

In this complex syndrome, management and treatment are complicated and delicate. First, 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 essential for a correct management of the case. Either the treatment either can by extraction of the SMMCI with space closure or by space opening associated to a prosthodontic restoration for the maxillary central incisor with an implant borne crown or a resin bonded bridge. Thus, an early orthodontic evaluation for an appropriate planning of the treatment timing is encouraged.

4. 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.

Author Contributions

ChB and FC performed the initial examination and collected clinical data. ChB provided the clinical dental care of the patient and continued performing regular clinical and radiographic follow-up. FM, FM, and AB supervised the clinical dental care of the patient. ChB and FC was responsible for the literature search and wrote the paper. FM revised and edited the manuscript and figures. HG provided comprehensive judgment and assisted in editing the final version of the manuscript. All authors read and approved the final version of the manuscript prior to submission

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We thank all clinical staff who participated in the treatment of the patient.

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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