INTRODUCTION

The presence of solitary median maxillary central incisor (SMMCI) is a rare anomaly of craniofacial development that can be associated with other anomalies of median line, being holoproscephaly (HPE) a form of lower expression (Johnson et al., 2008; DiBiase et al., 2008; Cho et al., 2006; Yang et al., 2005; Parenti & Perissutti, 2003). The SMMCI has an hereditary autosomal dominant transmission with incomplete penetrance and a high variable phenotype in familial cases (Tabatabaie et al., 2008; Bolan et al., 2009).

HPE has a prevalence of 1:10,000 (Koer et al., 2001). (DiBiase et al., 2006; Dubourg et al., 2007). The most severe cases of HPE are not compatible with life and often lead to miscarriages, while the milder forms (microforms) may simply manifest itself by the presence of a single maxillary central incisor (Hall et al., 2006; Dubourg et al., 2007; Hehr et al., 2004). Generally all patients with HPE have SMMCI, but not all patients with SMMCI present the HPE clinical features (Bolan et al., 2009).

The SMMCI could be a sign of a developmental disorder associated with variations in size and shape of neuregulin (Nann et al., 2001; Bolan et al., 2009). In most cases the patients with SMMCI have short stature and sometimes show endocrine disorders (Dubourg et al., 2007; Van Dijk et al., 2011).

The aim of this study is to present a clinical case and the treatment planning of a male patient with SMMCI, evaluated by Orofacial Genetic Department and Orthodontics Department of the Faculty of Dental Medicine, University of Oporto (FMDUP).

MATERIALS AND METHODS

SOLITARY CENTRAL INCISOR – CRANIOFACIAL EVALUATION AND TREATMENT PLANNING

The patient was studied following a protocol that including clinical examination, radiographic and photographic records and mounted models in a semi-adjustable articulator. Auxilary diagnostic data was requested consisting of analytical and imaging exams. It was additionally performed a literature research in PUBMED with the keywords “solitary median maxillary central incisor”, limited to Portuguese, English, French and Spanish idioms from 2000 to 2010. From this research 24 articles were obtained and 18 of them were included, associated with the theme.

CASE REPORT/RESULTS

The patient RHLR, caucasian male, aged 13 years and 9 months, came to the Orofacial Genetics department of FMDUP, with the task of improve the anterior dental esthetic (Figure 1). The case was evaluated and studied by the Orofacial Genetic and Orthodontic departments and the clinical diagnosis of SMMCI was assigned.

The facial frontal analysis revealed that both, middle and lower thirds of the face, were increased and although the lower third was slightly enlarged (Figure 2). Frontal examination highlights the following features: narrow but long face, broad nose, hypotelorism and convergent strabismus (Figure 2). The lateral facial analysis allowed us to verify that the patient had a straight profile, an advancement of the chin, a closed nasolabial angle and a facial asymmetry with the left ear positioned more anteriorly (Figure 2). The oral examination and the panoramic radiograph revealed the presence of a single median maxillary central incisor (the crown appeared to have a symmetrical shape) (Figure 2).

Figure 3 – Intraoral photographs.

The mandibular plane was very steep, leading to an important hyperteloropathy (Figure 3). The oral examination and the panoramic radiograph revealed the presence of a single maxillary median central incisor (the crown appeared to have a symmetrical shape) and the absence of the 45 tooth in the mandibular arch, which was found to be impacted (Figures 1, 5). After this examination, models were analyzed and a cephalometric study was performed. It was identified a Class III occlusion, fitted in a skeletal Class III type, followed by a retrogнатic and hypoplastic maxilla (Figures 3, 4, 7). The requested analytical blood test revealed a significant increase in the levels of the progesterone hormone (3.36 nmol/l, instead of the reference values of 0.35-0.83 nmol/l).

Figure 4 – Initial lateral and frontal X-ray with tracing.

From the orthodontic point of view and according to the literature about the treatment of SMMCI (Kjape et al., 2001), bolan et al., 2009). The SMMCI could be a sign of a developmental disorder associated with variations in size and shape of neuregulin (Nann et al., 2001; Bolan et al., 2009). In most cases the patients with SMMCI have short stature and sometimes show endocrine disorders (Dubourg et al., 2007; Van Dijk et al., 2011).

The oral examination and the panoramic radiograph revealed the presence of a single median maxillary central incisor (the crown appeared to have a symmetrical shape) (Figure 2).

Figure 5 – Panoramic radiograph.

The oral examination and the panoramic radiograph revealed the presence of a single maxillary median central incisor (the crown appeared to have a symmetrical shape) and the absence of the 45 tooth in the mandibular arch, which was found to be impacted (Figures 1, 5). After this examination, models were analyzed and a cephalometric study was performed. It was identified a Class III occlusion, fitted in a skeletal Class III type, followed by a retrogнатic and hypoplastic maxilla (Figures 3, 4, 7). The requested analytical blood test revealed a significant increase in the levels of the progesterone hormone (3.36 nmol/l, instead of the reference values of 0.35-0.83 nmol/l).

From the orthodontic point of view and according to the literature about the treatment of SMMCI (Tabatabaie et al., 2008; Bolan et al., 2010), the absence of the intermaxillary sutures and part of the intermaxillary suture, which is particularly evident on the occlusal upper photography as well as on the computerized tomography (Figure 1, 6). This clinical feature contradicts palatal transverse orthopedic disjunction (Figures 1, 6). In some cases it may be indicated extraction of the single central incisor and the promotion of mesial movement of the lateral incisors. From the periodontal standpoint, the patient had a generalized gingival inflammation. By clinical examination and using a computer-integrated system for periodontal probing (Florida Probe®, Florida Probe Corporation, Gainesville, USA) was diagnosed a mild chronic generalized periodontitis (Figure 8). From the 26 teeth evaluated, 2 sites (1%) of the 156 showed a probing depth more than 3.4 mm. 22 sites evidenced bleeding (14%) and suppuration or recession were not found. According to the multidisciplinary assessment and the treatment planning established, this included the stabilization of periodontal disease, followed by orthodontic treatment combined with orthognathic surgery, comprising an initial surgical palatal disjunction pursued by a maxillary advancement and mentoplasty.

Figure 6 – Intraoral model reconstruction by CT Scan.

CONCLUSIONS

The presence of SMMCI should not be regarded as a simple dental anomaly of development. It requires a medical evaluation performed by a multidisciplinary team, in order to establish the most appropriate treatment planning for the patient.

REFERENCES


Lertsirivorakul J, Hall RK. Solitary median maxillary central incisor syndrome occurring together with oromandibular-limb hypogenesis syndrome type 1: a case report of this previously unreported syndrome. Orphanet J Rare Dis. 2006;9:1-12.


Van Dijk et al. 2011.

Dubourg et al. 2007.

Cho et al. 2007;

DiBiase et al. 2006.

Cho et al. 2007;

DiBiase et al. 2006.

DiBiase et al. 2006.


Van Dijk et al. 2011.

Dubourg et al. 2007.

Cho et al. 2007;

DiBiase et al. 2006.