Journal section: Odontostomatology for the disabled or special patients *doi:10.4317/medoral.15.e859*
Publication Types: Review

Dental Treatment of Marfan Syndrome. With regard to a case

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Morales-Chávez MC, Rodríguez-López MV. Dental Treatment of Marfan Syndrome. With regard to a case. Med Oral Patol Oral Cir Bucal. 2010 Nov 1;15 (6):e859-62.
http://www.medicinaoral.com/medoralfree01/v15i6/medoralv15i6p859.pdf

Received: 28/02/2010
Accepted: 27/06/2010

Article Number: 16907 *http://www.medicinaoral.com/*

Abstract

Marfan syndrome is the most common dominant autosomic genetic disorder of the connective tissue. It has a reported incidence of 1 per each 5000 individuals without any distinction of gender or ethnicity. This pathology's diagnosis is mainly based on physical characteristics, presenting three main different symptomatic charts: neonatal Marfan, infant Marfan and classical Marfan. The mayor characteristic of these patients consists of an exaggerated length of the upper and lower limbs, hyperlaxity, scoliosis, alterations in the cardiovascular and pulmonary systems and atypical bone overgrowth. The individual implied in the present investigation concerned to a 14 year old male patient presenting multiple mouth lesions and dental alterations, attended in the Department of Pediatric Dentistry degree at the Dentistry School in the Santa Maria University. The patient has been treated following the necessary considerations required according to his systemic compromise d under oral premedication for decrease the anxiety and make easear the behavior management. The patirnt with MS has multiple oral decrease that may be diagnoticated a treated on time to increase the life quality of the patient.

Key words: *Marfan syndrome, dental management, oral premedication, patients with special needs.*

Introduction

Marfan Syndrome (MS) was firstly described by the pediatrician Antoine Bernard-Jean Marfan who reported an out of proportioned length of the lower limbs and fingers (1). The MS is a dominant autosomic genetic disorder which affects the elastic fibers of the connective tissue, showing itself in those systems/organs holding it, such as the cardiovascular, skeletal, dura mater, ocular, skin, teguments and lung (2, 3). This is mainly caused of mutations in the fibrilin glycoprotein's codified gene -1 located in the chromosome 15q21 (1,4). It has been described that the normal fibrilin inhibits the growth of the long bones and elastic fibers, through its tension control the growth of these, because of it, at these structures being altered, an exaggerated bone overgrowth is produced that better characterizes this

decease (2). This condition represents an incidence of 1 per each 5000/9800 individuals without any gender or ethnic distinction (1,-4).

MS is a multi-systemic disorder with typical manifestations which affect the skeletal, cardiovascular and ocular systems. On a skeletal level, an out of proportioned overgrowth of the long bones is observed which is frequently considered to be the most highlighted and evident feature. Nevertheless, other signs like pectum scavatum, scoliosis, articular hipermovility and flat foot may be presented. The cardiovascular pathology most frequently presented is the dilatation of the ascending aorta on the aortic sinuses level. These lesions constitute the main cause of mobility and mortality for patients with MS. The ocular system has been shown to be generally affected with a dislocation of the lens, however, several other pathologies can be developed such as cataracts or glaucoma (3,4).

A narrow cranium is present on the craniofacial area, with dolichocephaly features, deep palatal, jammed teeth, retrognathia or micrognathia, flat molars and descendant palpebral fissures. This type of paladar may cause a posterior cross bite. Also, the maxilar hypoplasia generally cause dental crowded. Westling et al. raise that crowded teeth is due to an increased overjet or an open bite (5,6). Dental structures may have hipoplasic stains with a higher prevalence than the rest of the population. This enamel defects plus the higiene deficit increase the caries incidence on these patients. It´s commun the roots may have distortion, pulpoliths and pulp obliterations. Bauss et al. (7) evaluated 21 rx of patients with MS and determinated that 20,7% presented pulpoliths and 7,9% pulpar obliterations. These anomalies may be considered at the time of endodontics treatment (7,8). The gingival and calculus index had a significant increased without many local irritants, with loss of gingival insertation and bone (9). Temporomandibular alterations are more prevalent because an articular deformation and ligament hiperlaxity. These damages may cause an articular block during a wide mouth open, pain during chewing or mouth opening click (10).

Clinical Case

A 14 year old male patient was treated in Department of Pediatric Dentistry degree at the Dentistry School in the Santa Maria University. At the time of questioning the patient's representative about his personal history(patient's), she argued that he is the youngest of 4 apparently healthy brothers, resulting from a controlled pregnancy, of which the parents were 34 and 36 years old at the time of gestation. With regard to the patient's medical background, his mother affirms that the young was diagnosed with Marfan Syndrome, dilatation in the aorta's base and subluxation of the crystals, plus, the teenager has been surgically intervened in three different occasions, because of 2 birth inguinal hernias at the time he was 9 months old, from tonsils and adenoids (3 years old) and an elongation of the calcaneus on the left foot respectively.

After the data collection corresponding the clinical history and the appropriate signature from the representative concerning to her informed consent, in compliance with the ethical rules of the institution, a clinical exam was carried out.

For the physical examination, a seize of 1,80 cm. range was determined from upper to average (Fig. 1) In the extra-oral examination, some syndrome-typical features were observed, such as an elongated/narrowed cranium or docichocephalism and very elongated limbs. Intra-orally, a very deep palatal was observed, Class II molar relationship by Angle and lack of space for all of the dental structures for which the incisive upper laterals were palatinized in relation to the centrals. Likewise, a presence of a generalized gingivitis associated to the dentobacterial plate and cavities in the 1.6 and 2.6 was observed. Furthermore, on a radiographical level, radicular cysts with radicular dilacerations between the 4.5 and 4.6 and agenesia for the second and third lower molars were present (Fig. 2, 3).

The dental treatment was performed under local anestethic using a maximun of 2 carpuls with vasoconstrictor because of the cardiovascular pathology. It consisted in the placement of a resin in the 1.6, a realization of an endodontic treatment on the 2.6 level with a posterior re-establishment, tartrectomy and exodoncy of

Fig. 1. Physical Characteristics of Marfan Syndrome. The long extremities are evident.

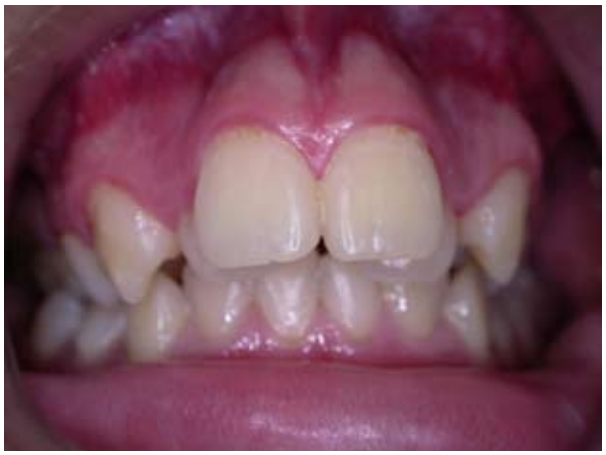

Fig. 2. Intraoral aspect. Front side. A gingivitis with dental plaque is observed. Also a severe malocclusion.

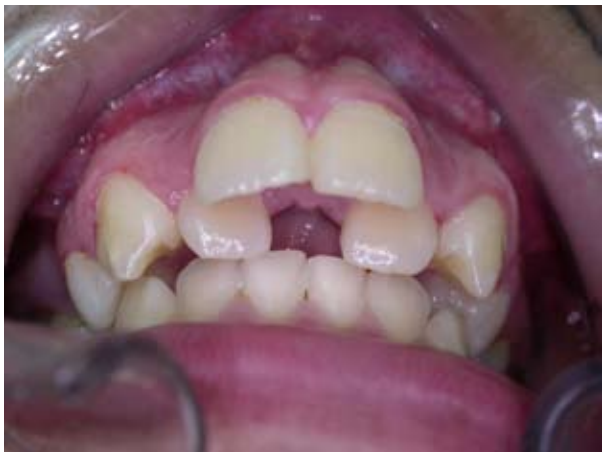

Fig. 3. Lateral incisor are not linning up because of the little transverse development of the maxilar.

the radicular rest. All of these procedures were carried out following a 2g amoxicillin antibiotic prophylaxis intake, which was taken one hour prior to the medical visit, due to the patient's cardiac pathology. The patient recieved an oral premedication of 5 mg diazepam the nigth before and one hour before the treatment to decrease anxiety. Subsequently, the patient was sent to the Orthodontic´s Department for the proper evaluation and occlusion's treatment.

Discussion

Marfan Syndrome is a dominant autosomic disorder of the connective tissue, as which Shiley et al affirm (1) presents a multisistemic affection as being part of one of the syndrome's main problems, the affectation of the skeletal system. On the other hand, typical skeletal characteristics of the syndrome such as the elongation of the extremities because of an exaggerated overgrowth of the long bones are observed in this patient. Concerning cardiac alterations, the most frequent, reported by authors such as Ammash et al (3) and Dean (4) is the dilatation of the ascending aorta on the aortic valve level. Taking into consideration the cardiac pathology of these kind of patients, it is imperative that preceding the performance of any odontological treatment implying bleeding, an application of the antibiotic prophylaxis by means of the intake of 2 grams of Amoxicillin one hour prior to the procedure or 600 mg of Clindamycin in case of allergic patients has to be carried out (11, 12). Nevertheless in the cases which required a long treatment is better to performed the dental treatment under sedation to use antibiotic prophilaxis just one time. Equally, some considerations must be taken at the time of selecting the proper anesthetics, because of the fact that authors such as Hirota and cols (12) establish that the epinephrine in these patients is capable of producing both an acceleration of the cardiac function and an increase in the cardiac output. It is also very common in these patients the presence of inguinal hernias, as observed in the exposed case, which had two.

On a craniofacial level, the palatal of the Marfan Syndrome is deep and stretch and both jawbones present retrognathia. Westilng and cols (5) studied 76 patient with this syndrome, observing in the 70% of them a dental collapse and an excessive increase of the over-jet, due to the minor jawbone development. These features are observed on a same manner in the present case. Related to the dental characteristics, De Coster and cols (8) report that in one population of 23 patients with MS, the majority presented a mayor risk of cavities and also a very high difficulty of treatment, because the existence of enamel hypoplasia, radicular deformities, and abnormal form of the pulp chamber. In the same way, they establish that the periodontal disease is presented with a higher frequency and severity in these patients. In the presented case, the patient had a generalized gingivitis mainly associated to plaque. As previously described in other reports, the patient presented a dilacerated radicular rest. No obliteration was found when performing endodontic treatment (7,8). Regarding to Temporomandibular Alterations, Bauss O et al (13) reported a prevalence of51.6% of articular disfunction and 24.2% of subluxation. (Table 1).

Patients with MS present endless medical compromises and mouth alterations that difficulty the overall dental treatment. Because of its medical conditions, a mayor predisposition to develop dental cavities, periodontal deceases and malocclusions are created. Early diagnoses of both dental and craniofacial anomalies, as well as an opportune appliance of an adequate treatment, could definitely develop a satisfactory prognosis of these type of patients, considerably improving their life quality.

Tabe 1. Oral Manifestation in Marfan Syndrome.

Authors	Oral Manifestations
Westing L, Mohlin B, Bresin A; 1998 (5)	Reported deep palate and maxilar and mandibular retrognatia in 70% of the evaluated patients.
De Coster PJ, Martens LC, De Paepe A; 2002 (8)	Reported a higher caries prevalence in patients from 0 to 17 years. High prevalence of hipoplasic stains. Radicular deformation and pulp obliteration. High gingival index and calculus
Straub A, Grahame R, Scully C, Tonetti M; 2002 (9)	Reported a case of a 41 years old patient with severe periodontitis, 5.6 mm of insertion loss, bone loss and dental mobility.
Bauss O, Sadat-Khonsari R, Fenske C, Engelke W; 2004 (13)	High prevalence of temporomandibular disfunction, subluxation and anterior displacement of articular disk in 21 patients with MS.
Bauss O, Neter D, Rahman A; 2008 (7)	High prevalence of pulpolitos and pulp obliteration in patients with MS.
Utreja A, Evans CA; 2009 (6)	Described 2 patients with severe Periodontitis without local factors.

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