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Clinical case

Artrogryposis multiplex congenita associated with intraoral changes – Multidisciplinary approach

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ABSTRACT

This article presents the clinical case of 21 years old female patient reporting history of Arthrogryposis Multiplex Congenita (AMC). The extraoral examination disclosed clinical AMC pathognomonic signs. The intraoral examination revealed slight compression of the maxillary arch, conical upper lateral incisors, absence of 17 and 35, 53 persistence, upper cuspids inclusion and agenesis of third molars. Additionally, there was a severe generalized shortening of the tooth roots, with a general 1:1 root/crown proportion. The extraction of 23 was planned due to its maxillary position. Relatively to the 13, the orthodontical traction with a microimplant was the option. Treatment planning established orthodontics to restore esthetics and function followed by rehabilitation with implants. Finally, the esthetic composite restorations of 12 and 22 were programmed, given the limited prognosis presented by fixed prosthesis in the cases of root/crown low proportions.

Artrogripose Múltipla Congénita associada a alterações – intraorais abordagem multidisciplinar

Palavras-chave:
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RESUMO

Este artigo apresenta um caso clínico de uma paciente de 21 anos, do sexo feminino, que relatou história de Artrogripose Múltipla Congénita (AMC). O exame extraoral revelou sinais clínicos patognomónicos de AMC. O exame intraoral revelou compressão da arcada maxilar, incisivos laterais superiores conoides, ausência do 17 e do 35, persistência do 53, inclusão dos caninos superiores e agenesia dos terceiros molares. Adicionalmente, verificou-se um encurtamento radicular severo generalizado, com a maioria das proporções raís/coroa 1:1. Foi planeada a extracção do 23, dada a sua posição na maxila. Relativamente ao 13, a tração ortodôntica com um microimplante foi a opção eleita. Planeou-se o recurso

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à ortodontia para restabelecer estética e função, seguida de reabilitação com implantes. Finalmente, planearam-se restaurações estéticas a compósito nos dentes 12 e 22, dado o limitado prognóstico oferecido pela prótese fixa em casos de baixas proporções raíz/coroa.

Introduction

Arthrogryposis Multiplex Congenita (AMC) is a rare congenital disorder,[1] affecting 1 in 3000[2–4] to 1 in 12,000[5] newborns. It involves the presence of multiple non-progressive,[1,3] symmetric joint contractures,[1,3,4] sometimes associated with muscle weakness and fibrosis.[1]

This disease can present an isolated form or it can also be associated with other congenital anomalies, as part of a syndrome, with or without central nervous system involvement.[6]

This condition's etiology is considered multifactorial and may be presented as a monogenic disease (autosomal recessive transmission, autosomal dominant or associated with the X chromosome), as a chromosomal disorder or as a congenital malformation (involving various organs).[7] AMC may also be associated with environmental factors such as infections, drugs administration, trauma, chronic diseases, oligohydramnios or abnormal uterus structure (affecting the mother and the developing fetus).[7] These factors described in the etiology of AMC are also common to approximately 7% of the congenital abnormalities in general.[3]

In the recognition of early clinical signs of AMC, in the last months of pregnancy, decreased fetal movement (fetal akinesia) is considered a common denominator to all AMC affected individuals, conditioning a variety of minor fetal deformities. It is important to notice the absence of movement, essential for joints and periarticular tissues development, leads to an increase of connective tissue around the immobilized joint with rippling of the skin covering the joint, muscle atrophy and changes in the joint surface depending on the position of the immobilization.[3,8]

The involvement of the temporomandibularjoint (TMJ) is a common AMC complication, conditioning the mandibular kinetics.[2,9] Other common features include the presence of micrognathia,[2,8] slightly shortened limbs, intrauterine growth restriction, pulmonary hypoplasia and short and/or immature bowel.[8] Some cases of AMC were also found associated with the presence of upper lateral conoid incisors,[10] hypodontia[11] and delayed tooth eruption.[2]

This paper focused on the presentation of a clinical case of AMC, emphasizing oral and craniofacial abnormalities and proposing a treatment approach.

Case report

A female patient, 21 years old, attended a dental appointment to assess orthodontic treatment need, referring the closure of existing dental gaps in the upper anterior arch as a priority. During the anamnesis, the patient reported an AMC history, diagnosed since childhood.

The extraoral clinical examination (Fig. 1) revealed pathognomonic clinical signs of AMC: multiple joint contractures, short stature, low set ears and dysplasia of the fingernails and toenails deployment. The analysis stressed a thin hypertonic upper lip and a low smile line TMJ clinical examination (by palpation, auscultation and mandibular kinetics evaluation) discarded clinical signs of temporomandibular disorder. The intraoral clinical examination (Fig. 2) showed a slight compression of the maxillary arch with anterior cross bite on teeth 12 and 22, Class II molar, decreased vertical overbite, upper lateral conoid incisors, absence of 18, 17, 13, 23, 28, 38, 35 and 48 and persistence of 53. In order to assess dental arch discrepancy, Bolton analysis[12] revealed a discrepancy with excess on lower anterior arch (Fig. 3).

Panoramic X-ray (Fig. 4A) complemented by a retroalveolar X-rays *status* (Fig. 4B) revealed the inclusion of upper cuspid, agenesis of third molars (upper and lower), absence of 17 and 45 and a generalized severe dental root shortening.

Ricketts cephalometric analysis (Fig. 4C and D) revealed a skeletal Class II, with a retro and micrognatic mandible, an orthopositioned maxilla, a mesocephalic facial type, proclined and orthopositioned upper and lower incisors, a decreased interincisal angle and a lip retraction. Functional analysis according to Multifunction System (MFS) classification[13] showed type 1 nasal collapse (narrow nostrils without collapse), type 2 adenoids (slightly convex), type 2 tonsils (appear slightly), normal swallowing, nasal breathing, type 1 tongue mobility level (tongue touches the palate). In order to improve

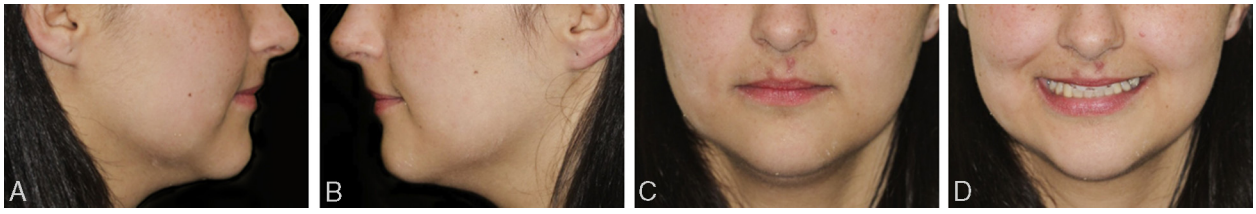

Fig. 1 – Facial appearance at rest – (A) right side, (B) left side, (C) front, (D) smile.