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Cleidocranial dysostosis - case report of a multidisciplinary approach

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Resumo

A disostose cleidocraniana é uma síndrome genética rara, autossómica dominante. Neste caso particular, os muitos desafios dos tratamentos foram resolvidos por uma equipe odontológica, cujo traccionamento ortodôntico de dentes inclusos associados com a cirurgia ortognática bimaxilar foi o principal foco. O objectivo deste tratamento ortodôntico-cirúrgico foi restaurar a oclusão funcional e melhorar a estética facial, aproveitando a vantagem de todos os dentes impactados que caracterizam esta síndrome.

Palavras-chave: Disostose cleidocraniana; tratamento ortodôntico; cirurgia ortognática.

Abstract

Cleidocranial dysostosis (CCD) is a rare genetic syndrome, autosomal dominant. In this paper a patient with cleidocranial dysostosis was treated under a multidisciplinary basis. Many treatment challenges were solved by a dental team, in which the orthodontic traction of unerupted teeth associated with bimaxilar surgery had the main focus. The aim of this surgical-orthodontic treatment was to restore the functional occlusion and to improve the facial aesthetics, taking advantage of all the impacted teeth which characterize this syndrome.

Keywords: Cleidocranial dysostosis; orthodontic treatment; orthognatic surgery.

INTRODUCTION

Cleidocranial dysostosis (CCD) is a rare genetic syndrome, autosomal dominant, recognized by Marie and Sainton in 1898[1]. It is associated with a spontaneous mutation in the RUNX2[2,3] gene also identified as Core-binding factor 1 (Cbfa1)[1,4,5] which is essencial for osteoblast and dental cell differentiation[2]. This responsible gene, located in the short arm of chromosome 6, was identified in 1997[4]. It is responsible for osteoblast formation, chondrocyte differentiation and vascular invasion of calcified cartilage[1]. The phenotype shows abnormal clavicles (hypoplasia or aplasia of clavicules)[4-6] wormian bones and supernumerary teeth[2]. Retarded ossification in bone precursors, especially at junctions is the result of early developmental disorder of mesenchyme or connective tissue and can lead to defective portions of skeletal structures[2]. The principal affected bones (intramembranous ossification) are the cranial vault, clavicles, maxilla, nasal and lacrimal bones[7]. The prevalence of CCD is one per million, displaying no sex or ethnic group predilection[5].

This syndrome can be diagnosed by prenatal ultrasonography, but usually it is detected later due to dental problems or other not related medical pathologies[1,8] (Table 1).

In this paper a patient with cleidocranial dysostosis was treated under a multidisciplinary basis.

CASE REPORT

A 18-year-old female patient had a complaint of chewing difficulty. She presented with an asymmetric braquifacial appearance, a concave profile and a low smile line. The intra-oral examination showed a V shaped palatal arch, the presence of deciduous canines and the absence of twelve permanent teeth. The patient had a short stature and an edentulous and aged facial appearance (Figure 1). A panoramic radiograph and lateral cephalogram showed a delayed eruption of permanent teeth

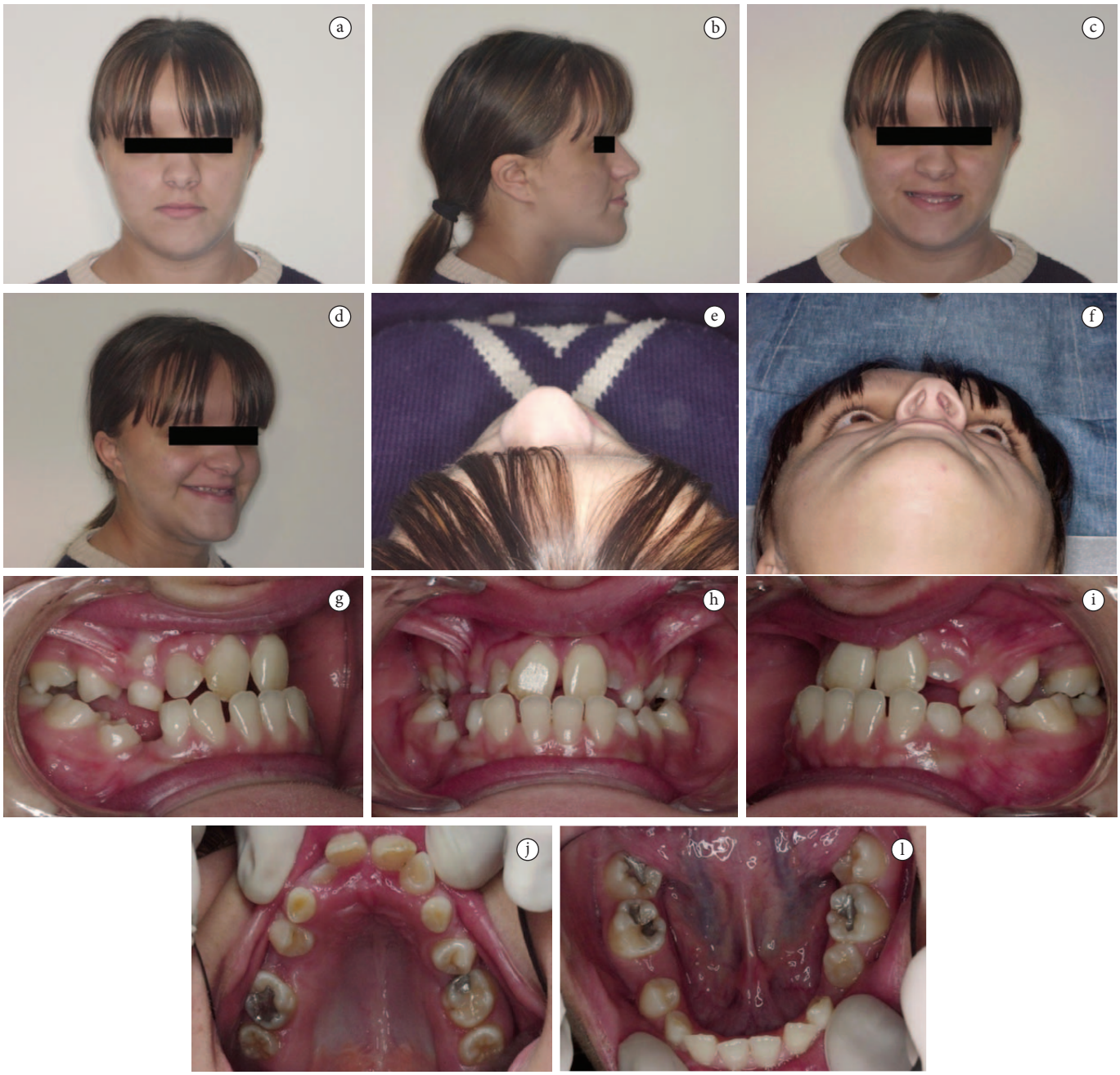

Figure 1. a-l) Extra-oral and intra-oral pretreatment photos.

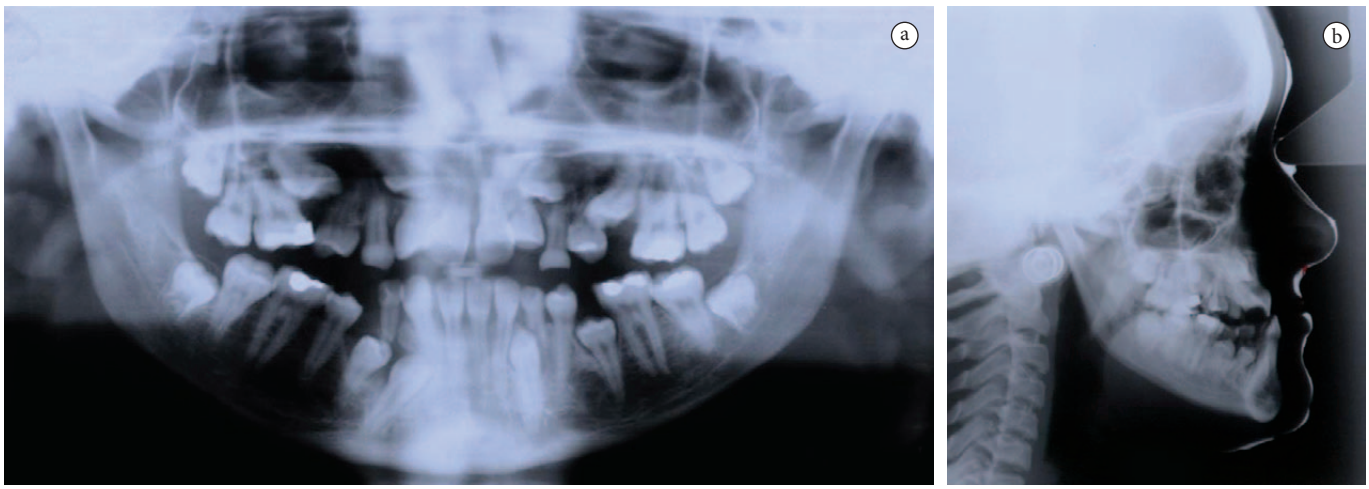

Figure 2. a) Panoramic radiograph; b) lateral cephalogram.

with a generalized hypercementosis, hypoplastic midfacial bones, and an obtuse mandibular gonial angle with a skeletal class III relationship (Figure 2). The diagnosis of CCD was made and confirmed with the aplasia of clavicles confirmation (Figure 3).

An orthodontic treatment with surgical exposure of unerupted teeth followed by orthodontic traction previously to an orthognatic surgery (Le Fort I Osteotomy with Bilateral Sagittal Split Osteotomy - BSSO) was the treatment plan (Figure 4 and 5).

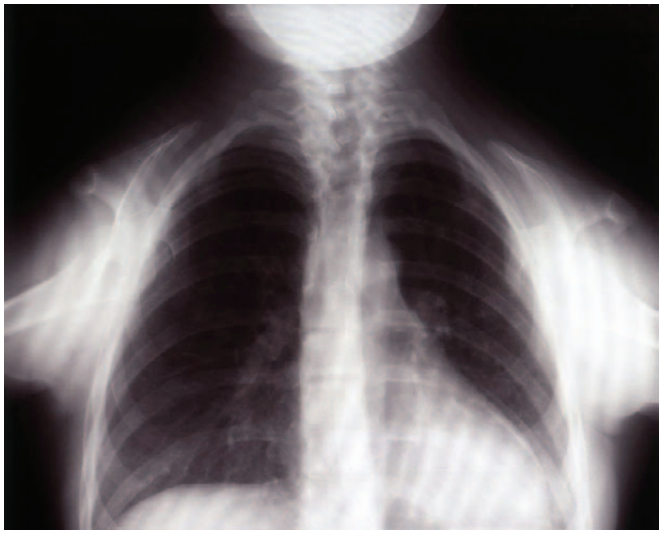

Figure 3. Chest radiograph. Note the aplasia of clavicles.

Table 1. Cleidocranial dysostosis – main characteristics[8]

Pathology	Hypoplastic/normal bones
Clinical features	
Height	Normal
Skull vault	Bigger (brachycephalic)
Skull base	Normal
Diffuse sclerosis	Absent
Cranial sutures & fontanelle	Open
Wormian bones	Present
Maxilla	Hypoplastic
Maxillary sinus	Under-pneumatized
Gonial angle	Normal
Clavicle	Aplastic/hypoplastic
Hands & feet	Normal
Tendency for fracture	Normal
Long bones	Normal
Bone texture	Normal
Cranial nerves	Not involved
Extra-medullary hemopoiesis	Absent

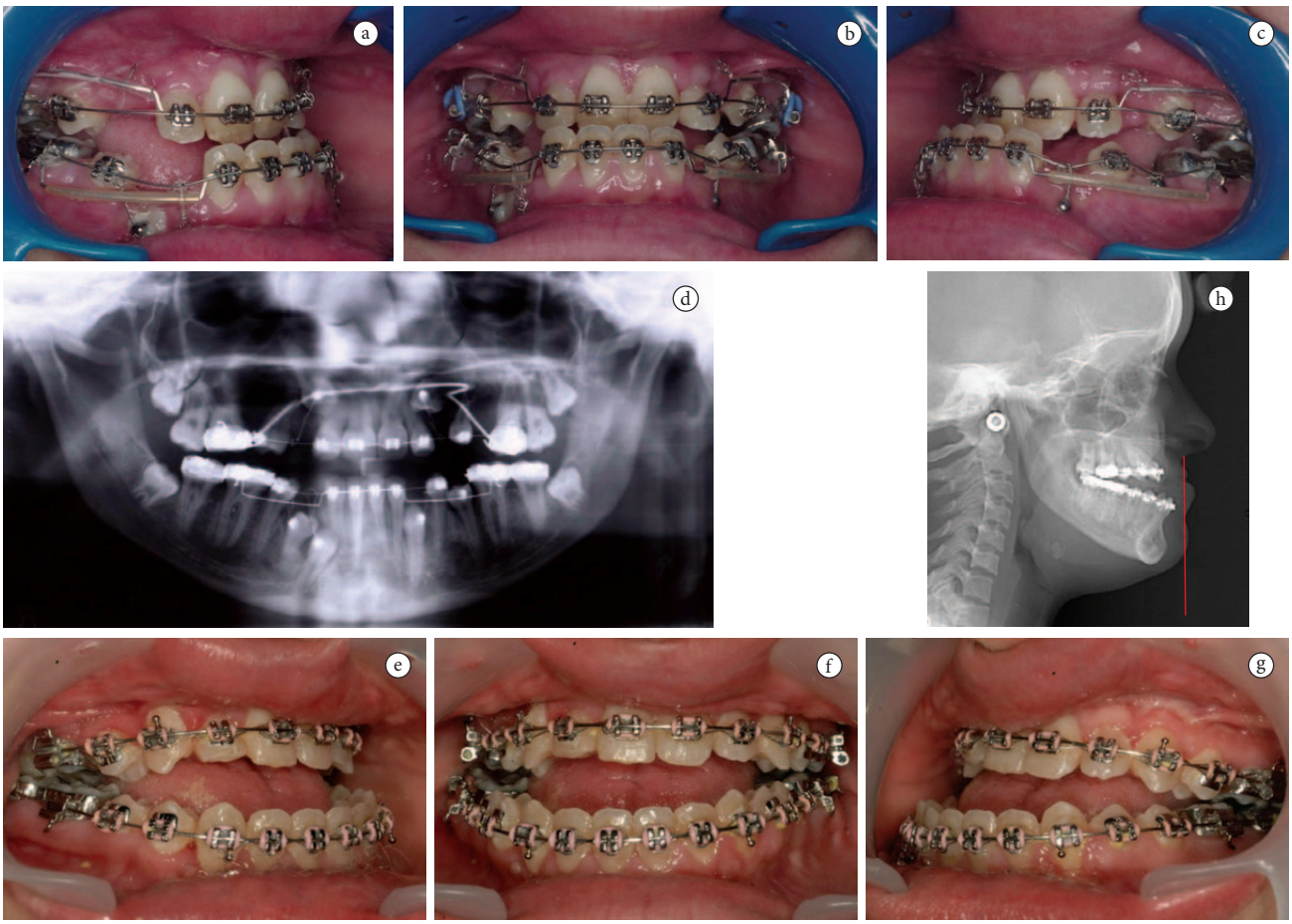

Figure 4. a-h) Presurgical radiographs and photos.

Figure 5. a-j) Final radiographs and photos.

DISCUSSION

Reviewing the literature considering patients with CCD, the concave profile indicates a skeletal class III with a hypoplastic maxila and a closing mandibular rotation, having a poor vertical alveolar growth[6]. The maxilla is short vertically but not antero-posterioly[9]. The panoramic radiograph is a valuable auxiliary exam to confirme CCD diagnosis. The characteristic dental abnormalities are supernumerary teeth, delayed eruption and impaction of permanent teeth, prolonged retention of deciduous teeth, and multiple impactions[2,4,6] as demonstrated in this case. The impaction can result from local biomechanical obstacles such as childhood maxillofacial or dentoalveolar trauma, reconstructive surgery of facial skeleton, malpositioning of adjacent teeth, thickened overlying osseous or mucosal tissues, low correlation between maxillofacial skeletal development and tooth maturation, direct or indirect effects of cysts or neoplasm[10], the increase in amount of acellular cementum of the roots of affected teeth instead of cellular cementum[2]. In this case, these biomechanicals obstacles were removed with surgery and orthodontic traction, using fixed appliances. At the same time, the second maxillary premolars were carefully extracted due to its abnormal morphology and malposition, probably related to the lack of space and blocked eruption[10]. Only a minimal amount of bone was removed to bond attachments

to enamel, in order to avoid the damage of bone support that would harm the periodontal prognosis of the tractioned teeth. The cephalometric values of normal individuals used to determine the labiolingual incisor position are inappropriate to patients with CCD. That is why clinical judgement is the main factor to plan the orthodontic treatment. Nevertheless, a long term retention is necessary[7].

After this 3 years interdisciplinary complex treatment, the patient had an acceptable facial appearance and an appropriate oral function.

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