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Orthodontic treatment of a patient with cleidocranial dysplasia: A case report

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Abstract

Cleidocranial dysplasia is a rare genetic disease with various forms of expression. It is mainly manifested by numerous and evolving skeletal, clavicular, and craniofacial anomalies. Among them, dental signs are still often at the origin of the diagnosis and are among the most disabling for the patients. Thus, in adolescence, the therapeutic approach focuses almost exclusively on the orofacial sphere and mobilizes all the ontological disciplines. Today, orthodontic treatment strategies take into account the pathophysiological mechanisms of the condition and lead to satisfactory results justifying long treatment periods.

Keywords: Cleidocranial Dysplasia, Dental Anomalies, Orthodontic Treatment

Introduction

The term cleidocranial dysplasia comes from the Greek words cleido (collar bone), kranion (head), and dysplasia (abnormal formation). Dysplasia refers to any abnormality in the development of a cell, tissue, or organ resulting in deformation or malformation of a cell, tissue, or organ. However, dysostosis refers to a deformity that preferentially affects one or more bone parts. Therefore, since 1970, the term "dysplasia" has been used rather than "dysostosis" which is a more restrictive term [1]. Cleidocranial dysplasia (CCD) is a rare autosomal dominant disorder of dysplasia and congenital bone malformation with an incidence of approximately 1 per million births [2].

Abnormalities are both qualitative and quantitative and are preferentially located in the medial skeletal segments. The disease was first reported in 1760 by Meckel [3] and was definitively renamed CCD by Marie and Saton [3] in 1897, attributing to it asymptomatic triad consisting of clavicular aplasia, cranial dystrophies, and hereditary transmission.

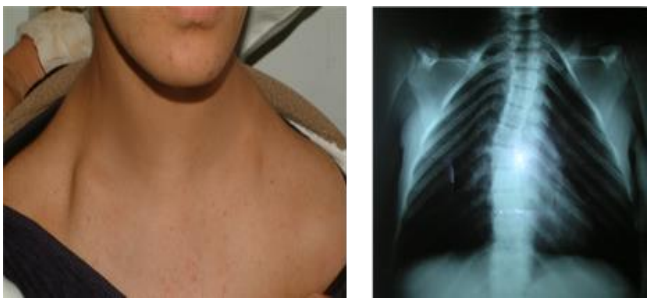
Its diagnosis can nowadays be genetic, possibly prenatal, but it remains mostly clinical and is still often based on the constant craniofacial and Bucco-dental disorders, which will remain the main concern of the patient with CCD [4].

Contrary to other syndromes associating skeletal and dental anomalies, the clinical picture of CCD is not well known and its orthodontic management remains a major challenge for orthodontists because of the absence of a global treatment plan applicable to all patients due to the extreme variety of clinical cases.

The purpose of this work is to report through a clinical case the orthodontic management of a patient suffering from CCD.

Cas clinique

A 25-year-old female patient, with no particular medical or surgical history, presented to the clinic with an anterior gap and dental crowding. The patient presented with a slightly subnormal weight and height development: narrow thorax, drooping shoulders, and developed musculature (fig 1a, fig 1b).



a b

Fig 1: a) Extraoral examination of the chest cause drooping shoulders, **b)** Chest x-ray showing a narrow chest

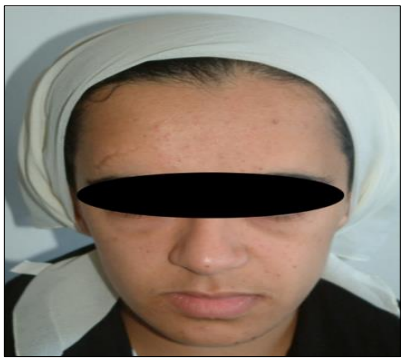
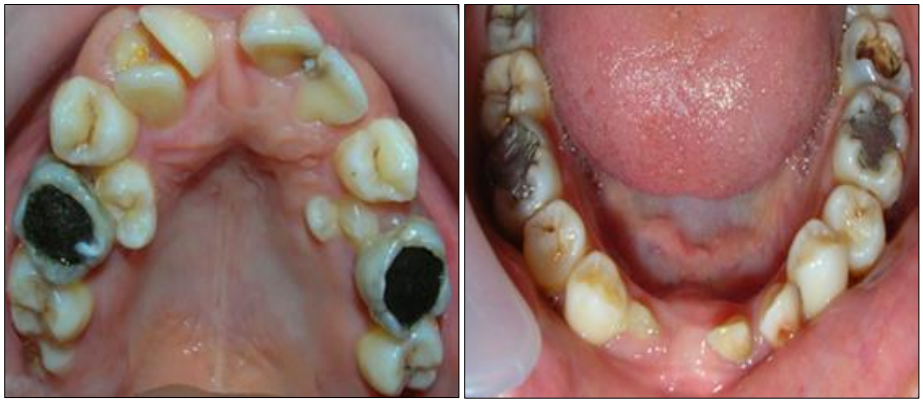


Fig 2: Maxillofacial examination showing a narrow triangular face

Maxillofacial examination revealed prominent frontal bumps with a more developed cranial mass in the transverse direction than in the sagittal direction (brachycephaly). The face was triangular and flat, the cheekbones erased, the nose was plane with an enlarged root and a septum slightly deviated to the right.
An eversion of the lower lip compared to the thinner upper lip was also noted (Fig 2).

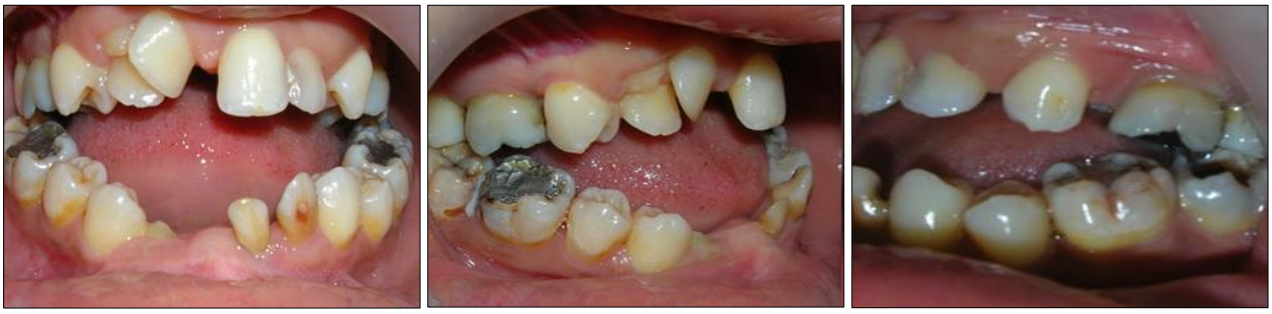
Endobuccal examination revealed maxillary endognathy, a narrow and deep palate, significant maxillary crowding with a palatal position of 15 and 25, absence of 13 and 23 on the arch, rotation of 11, and presence of caries on 22, and occlusal restoration on 16 and 26 (fig 3a). The mandibular arch showed a wide 'U' shape with a weak tongue and the presence of caries, the absence on the arcade of 33, 31, 41, 42, and 43. We also noted the persistence of deciduous teeth 71 and 81.



a b

Fig 3: a) Endobuccal view of the maxilla, **b)** Intraoral view of the mandible

The inter-arch examination in the front view showed a significant anterior-posterior gap with a bilateral posterior crossbite and lingual interposition (fig 4a). In lateral view, a right and left molar class 1 was noted, with the absence of canine landmarks for the right (fig 4b) and left (fig 4c) canine relationship.



a b c

Fig 4a: Interarch relationship in the front view, **Fig 4b:** Interarch relationship in the right profil view, **Fig 4c:** Interarch relationship in the left profil view

The initial panoramic radiograph revealed (Fig.5)

- Bone-wise:
- poorly developed maxillary sinuses,
- thin coronoid processes,
- the right condyle slightly narrower than the left
- Dental:
- anarchic dental axes (mesio or distoversion, rotation),
- persistence of temporary teeth (71 and 81),
- numerous retained teeth (13, 23, 28, 33, 31, 41 and 43) and agenesis (32 and 42).

- numerous occlusal restorations (16, 26, 37, 36, 46).

The patient's profile teleradiography showed the following signs: prominent frontal humps, absence of the nasal bone, mandibular pseudo-prognathism, open mandibular angle with an anteroposterior gap, and many impacted teeth (Fig.6).

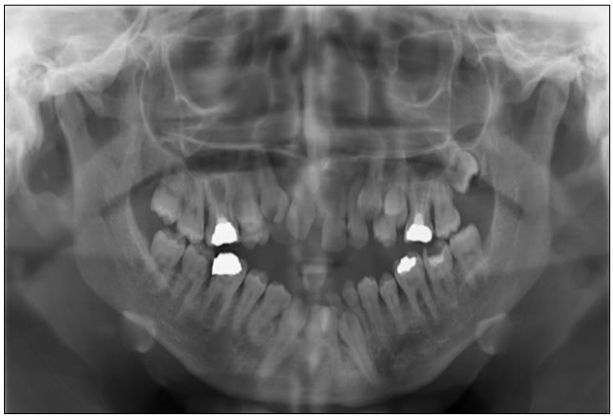


Fig 5: Panoramic X-ray

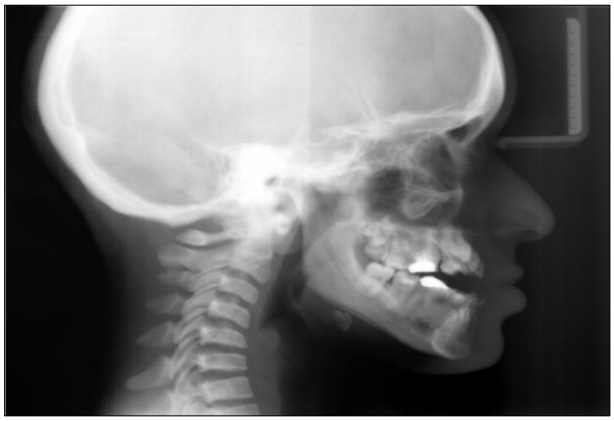
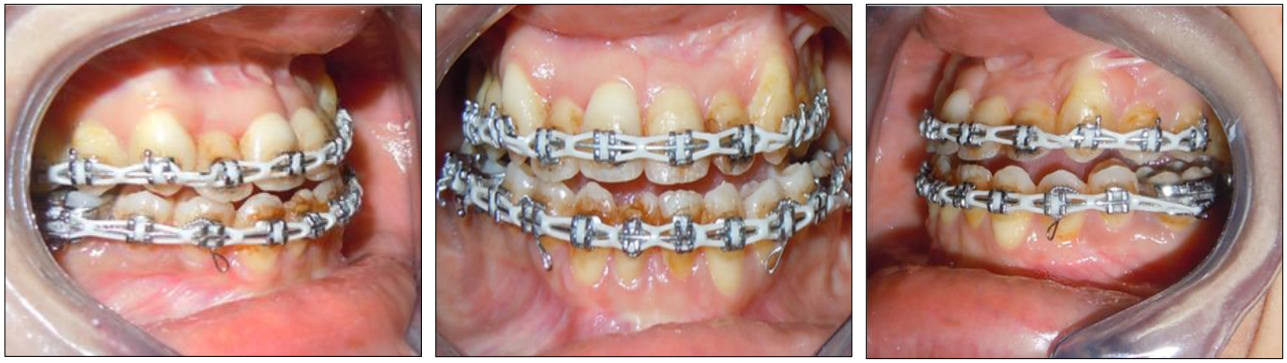


Fig 6: Profile teleradiography

Based on the clinical and radiographic findings of these examinations (clavicular involvement, dental anomalies, and craniofacial deformities), the diagnosis evoked was cleidocranial dysplasia. Orthodontic treatment was the preferred solution in this clinical case after the orthopedic phase (fig.6).



a b c

Fig 6: a) Orthodontic interarch relationship in the right profil view, **b)** Orthodontic interarch relationship in the front view, **c)** Orthodontic interarch relationship in the left profil view

Discussion

Cranial Cleido Dysplasia is a rare condition with an autosomal dominant mode of transmission. The risk of transmission is therefore 50%. Its penetrance is complete, which means that any person carrying the genetic anomaly is ill [1, 2]. The clinical pictures vary in number and intensity and over time. However, the description of the syndrome can be approached according to four cardinal signs: clavicular aplasia or hypoplasia, craniofacial deformities, anomalies of the dentition, and dentition such as malocclusions, delayed or absent exfoliation of temporary teeth, and multiple dental inclusions contrasting with pseudo-anodontia.

At present, it is difficult to establish a comprehensive treatment plan applicable to all patients [5]. The low eruption potential of the teeth and the presence of orthodontic anchorage problems is a real challenge for the orthodontist. Therefore, orthodontic treatment must be individualized since cases are extremely variable.

The direction of the treatment plan is largely dependent on early diagnosis and management. Early management during growth allows for transverse and sagittal development of the bony bases [5]. Thereafter, there is a range of intervention methods that allow a precise and individual choice according to the type of inclusion encountered; ranging from ortho-surgical treatment to extractions and prosthesis [6, 7].

In our case, orthodontic treatment was the preferred solution after the orthopedic phase. It consisted of performing post-orthodontic coroplast of the mandibular canines and first premolars into lateral incisors and lower canines, respectively, to correct the canine class III into canine class I. This choice is mainly guided by the inclusions of 13 and 23 and the agenesis of 32 and 42.

Conclusion

The dental physician can play a crucial role in both the detection and treatment of cleidocranial dysplasia. The rarity of the syndrome and the diversity of clinical manifestations imply a still very empirical approach in the therapeutic steps. Early treatment during growth allows for transverse and sagittal development of the bony bases and thus avoids heavy interventions in adulthood.

Conflict of interest

None

Références

1. Petropoulos VC, Balshi TJ, Wolfinger GJ, Balshi SF. Treatment of a patient with cleidocranial dysplasia using

a single-stage implant protocol. J Prosthod. 2011; 20(1):26-31.
2. Cooper SC, Flaitzi CM, Johnston DA. A natural history of cleidocranial dysplasia. Am J Med Genet. 2001; 104(1):1-6.
3. Oyer CE, Tatevosyants NG, Cortez SC, *et al*. Cleidocranial dysplasia with neonatal death due to central nervous system injury in utero: case report and literature review. Pediatr Dev Pathol. 1998; 1:314.
4. Jensen BL, Kreiborg S. Dental treatment strategies in cleidocranial dysplasia. Br Dent J. 1992; 172:243-247.
5. Zhang CY, Si Y, Wang XZ, Sun XY, Yan WJ, Zheng SG. Early dental treatments for patients with cleidocranial dysplasia. Chin J Dent Res. 2015; 18:51-57.
6. Lafargue H, Richard B, Gorry P, Marteau JM, Boileau MJ. La dysplasie cléido-crânienne : attitudes thérapeutiques et recommandations. À propos d'une observation clinique. Actualités Odonto Stomatologiques. 2009; 247:241-254.
7. Moukram K, Alaoui OM, Tahiri A, Et I Ben Yahya. La complexité de la prise en charge odontostomatologique de la dysplasie cléido-crânienne (DCC): Rapport de cas et revue de littérature. Med Buccale Chir Buccale. 2016; 22:297-303.