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Early Dental Approach of a Young Patient with Costello Syndrome: Case Report

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Case Report

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Abstract: Costello Syndrome (CS) is a rare congenital disease characterized by a wide range of cardiac, musculoskeletal, dermatological, developmental and dental abnormalities. Intraoral, enamel defects, delayed eruption and malocclusion, are the most common findings. We report the case of a three-year-old girl who was referred to the pediatric dentistry department of hospital la Rabta (Tunisia). It exhibited many of the dental manifestations of CS as well as general abnormalities. In our early treatment approach, the anterior teeth which presented enamel defect were restored with composite resin to adjust the incisor shape and improve esthetics and chewing function.

Keywords: Costello Syndrome, hereditary disease, syndrome, specific oral findings, dental management.

INTRODUCTION

Costello Syndrome (CS), a rare autosomal dominant disorder, is a multiple congenital anomaly disorder characterized by craniofacial malformations, dermatologic anomalies, cardiac defects, musculoskeletal abnormalities, growth delay and cognitive deficits [1, 6].

The estimated number of patients worldwide is 300. Estimated birth prevalence has been reported to be 1/300000 to 1/1.25 million [7]. It is caused by activated RAS/mitogen-activated protein kinase (MAPK) signalling. Specially, activating mutations in HRAS causes CS [3].

Among craniofacial feature common in CS, we can find macrocephaly, bitemporal narrowing, convex facial profile, full cheeks, short nose and large mouth [2].

CS patients have a characteristic dental phenotype that includes enamel hypo-mineralization, delayed tooth development and eruption, malocclusion, missing or supernumerary teeth, gingival hyperplasia, thickening of the alveolar ridge and high palate [5, 8].

This clinical observation exposes a rare case of Costello Syndrome in Tunisia (North Africa). It stresses oral manifestations and the importance of the pedodontist in the prevention of the complications of this pathology.

CASE PRESENTATION

A 3-year-old female patient was been referred from the pediatric service at the Rabta University hospital-Tunis-Tunisia to pediatric dentistry department in April 2017 for gingival hyperplasia and enamel defects.

Through clinical interview with the parents, we noticed that the patient was not born from a consanguineous marriage. The pregnancy was normal, without any special incidents. The child was born by caesarean section and the birth weight was 4500g. The three older brothers of the patient are healthy.

She was very ticklish, making physical examination quite difficult. She had coarse facial features and increased skin pigmentation. Her hair was light sparse and curly. She had a relative macrocephaly: a prominent forehead with low frontal hairline, a bitemporal narrowing and low-set-ears. She also displayed strabism, midfacial hypoplasia, ocular hypertelorism and a wide based nose with a depressed nasal rood, rounded tip and anteverted nostrils. Her nasal bridge was depressed. Her mouth was wide with prominent and full lips as shown in Figure (1A).

The patient has a delayed growth. Actually, her weight was 11 Kg and her height was 55 cm as presented in Figure (1B).

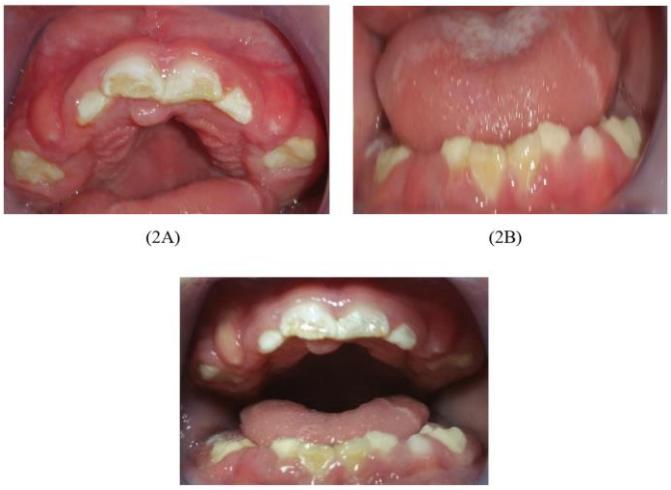
(1A) (1B)

Fig-1: specific facial features: macrocephaly, bitemporal narrowing, low-set-ears, strabism, midfacial hypoplasia, ocular hypertelorism and delayed growth

At this age, she is delayed in walking, in speech and vocabulary. The child has cardiac problem such as hypertrophic cardiomyopathy and pericarditis. She suffered also from hyperthyroidism.

Intraoral examination revealed enamel hypomineralization associated to enamel defect notably in the incisal edge of anterior teeth. A delayed tooth development and eruption is observed (Figure 2A, 2B).

We found also gingival hyperplasia, high palate with thickening of posterior maxillary alveolar ridge, heavy incisal wear on the mandibular and the maxillary central incisors and thickening of the anterior mandibular alveolar ridge (Figure 2A, 2B, 2C).



(2A) (2B)

(2C)

Fig-2: Intra oral examination: enamel defects, gingival hyperplasia, high palate with thickening of posterior maxillary alveolar ridge, heavy incisal wear and delayed eruption.

In our early treatment approach, the anterior teeth which presented enamel defect were restored with composite resin to adjust the incisor shape and improve esthetics and chewing function (Figure-3).

Gingivectomy is planned to be conducted in this case right after the eruption of all deciduous teeth

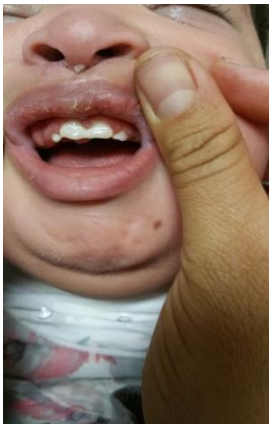


Fig-3: Restauration of incisor with composite resin

DISCUSSION

We report a 3-years-old young girl with Costello Syndrome, in addition to general feature, presented some specific oral abnormalities.

As many organ systems can be affected, treatment may require the coordinated efforts of a team of specialist.

The most common oral features in CS reported in the literature are delayed eruption, enamel defects, delayed development and high palate [5].

Curently, there is no cure for Costello syndrome although early intervention is important in alleviating symptoms and preventing complications. Treatment should be directed towards the specific needs of each individual and his or her family.

The enamel defects should be restaurated by acid-etched composite resin to ensure masticatory function, to protect teeth against further wear, sensitivity, and plaque accumulation and the patient's esthetic appearance.

CS individuals do not present with unique dental pathologies requiring specific treatment. Like the general population, patients with CS require routine dental examinations and appropriate hygiene and restorative care.

Careful oral hygiene instructions to patients and their families are necessary, since individuals with CS may not have meticulous oral hygiene habits. Some individuals with CS may be anxious dental patients due to cognitive delay and oral aversion, and thus, these individuals should be seen early and often by the dentist to accustom them to dental treatment [4, 8].

Although the caries incidence for this specific pathology was not strikingly high, CS individuals may be at higher risk due to hypomineralized enamel. Increased fluoride treatment, whether in office fluoride varnish or at home fluoride rinse besides to fluoride toothpaste, may be advised. In addition, enamel structure abnormalities may lead to coducted pathologic wear of the teeth due to bruxism, and for severe tooth abrasion, a custom mouthguard may be considered [5].

Delayed tooth development and eruption should be explained to patients and their families to alleviate any concerns. If there is a significant delay in eruption, panoramic X-rays are recommended to determine tooth development stage [8].

Furthermore, dentists should be aware and monitor the development of malocclusion in patients with CS and be prepared to refer these patients to an orthodontist for treatment if necessary [4].

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Conflict of Interest

There is no financial interest or any conflict of interest.

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