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(CASE REPORT)

Cerebral gigantism: sotos syndrome: A case report

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Abstract

Sotos syndrome is a genetic condition characterized by overgrowth, advanced bone age and a typical facial appearance. Oral findings include a high-arched palate, premature eruption of the teeth and dental agenesis. This paper documents a child diagnosed with Sotos Syndrome and describes the primary clinical features, the disease-specific, oral and dental findings, and dental care management of this patient.

Keywords: Sotos syndrome; Cerebral gigantism; Oral findings; Dental agenesis.

1. Introduction

Sotos syndrome or cerebral gigantism is a rare genetic disorder characterized by increase birthweight, excessive growth during the first few years of life and distinctive facial features. It is the result of abnormality of a single gene (NSD1) located on chromosome 5. Its prevalence is estimated to be between 1/10,000 and 1/50,000 [1].

The aim of this article is to describe the general physical characteristics and oral manifestations related to this rare syndrome.

2. Case report

A 10 year old male patient reported to the Department of Pediatric and Preventive Dentistry, Faculty of Dentistry, Rabat (Morroco), with the chief complaint of multiple decayed teeth.

Medical history when elicited revealed that the patient was born out of non-consanguineous marriage, full term ,delivered vaginally in hospital with no history of birth asphyxia .He has one elder sibling (sister) who is healthy. The family history revealed that the heights of the parents were normal and recorded as 1,65m m for mother and 1,80m for fater.

During her pregnancy, the 25-year-old mother was treated for easy preeclampsia, from week 25 of gestation. On follow-up ultrasounds, an abnormal increase in the cranial perimeter was seen.

Patient's weight at birth was 3, 3 Kg; however the height was not recorded. The neonatal period was marked by various complications: food intake difficulties, hypotonia and jaundice treated by phototherapy.

The patient has an early primary tooth eruption at only 2, 5 months. He began to speak when he was 5 years old and started walking at 8 months old.

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A detailed medical history revealed that the patient was having, asthma, scoliosis, urinary incontinence and asymmetry of the lower limbs.

The diagnosis of Sotos syndrome is established at 2 years old by identification of a deletion encompassing NSD1 on molecular genetic testing. The diagnosis was suspected because of the presence of the following characteristics:

- Characteristic facial appearance
- Learning disability
- Overgrowth

Clinical examination showed that the patient present a dysmorphic features assosiated with Sotos syndrome including: macrocrania, prominent forehead, long narrow face, pointed chin, downslanting palpebral fissures, flushing malar and receding hairline. (Fig.1, 2)

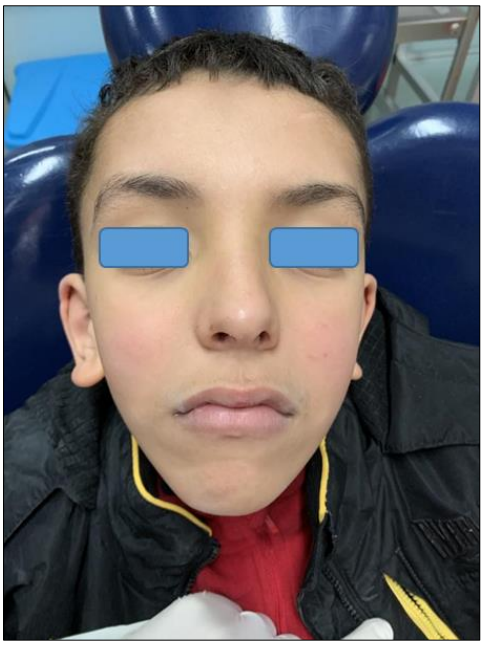

Figure 1 Extra oral photograph showing a prominent forehead, long narrow face, pointed chin, flushing malar and receding hairline

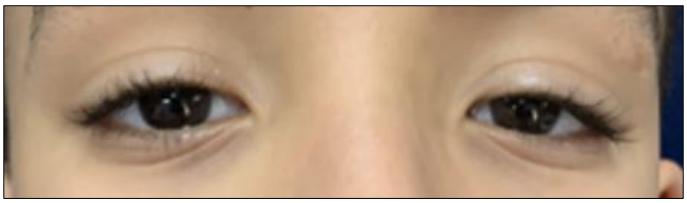

Figure 2 Extra oral photograph showing a downslanting palpebral fissures

He has been under psychologist's control because of behavioral problems like temper tantrums, inattention, hyper-activity, impulsiveness, poor concentration abilities and difficulty with peer group relationships.

Intra oral examination showed a poor oral hygiene, ogival palate, supragingival calculus and a multiple caries. (Fig.3)

Radiographic examination shows the existence of multiple agenesis of permanent teeth; 4 in the upper jaw and 4 in the lower jaw. (Fig.3)

Because of the child's uncooperative behavior, oral care was performed under general anesthesia. The dental treatment plan was based on the patient's caries risk and consisted of restoration of carious teeth and extraction of unrestorable teeth. As there were no systemic abnormalities, antibiotic prophylaxis was not recommended. Oral hygiene was improved through professional oral prophylaxis.

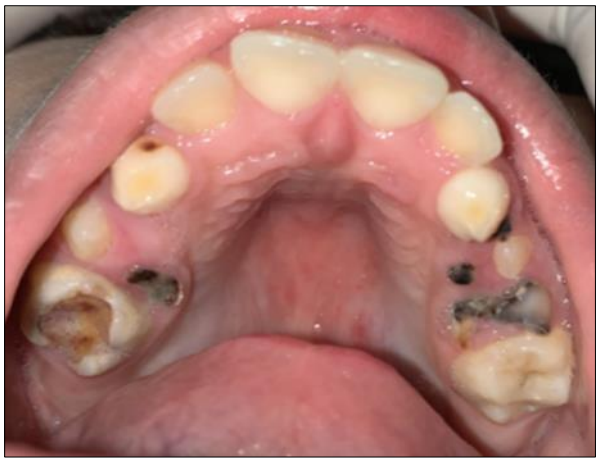

Figure 3 Intra oral photograph showing an ogival palate and a multiple caries

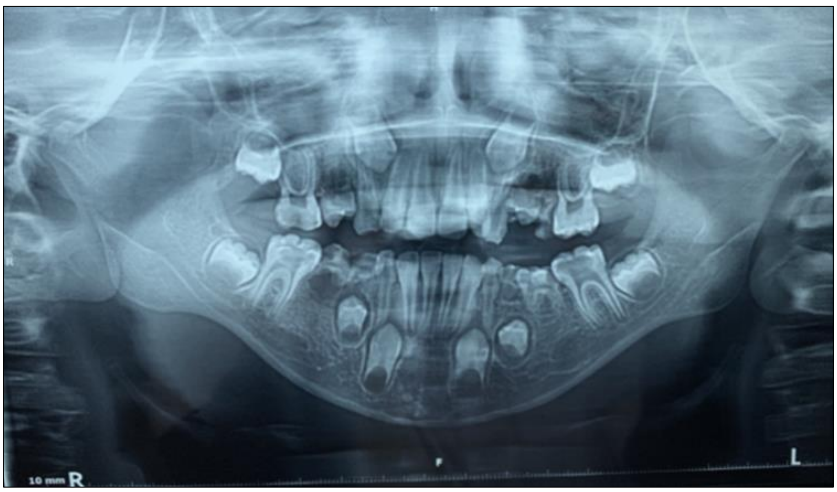

Figure 4 Panoramic X-ray showing the existence of multiple agenesis of permanents teeth (15, 18, 25, 28, 35,38, 45, 48)

Sotos syndrome is associated with multiple dental agenesis in 69% [2]. Therefore, pedodontists should be aware of this. Early diagnosis and interventions are necessary in order to preserve teeth as long as possible.

3. Conclusion

Patients with Sotos syndrome require a multidisciplinary approach with collaboration between different specialties. Preventive therapy is considered essential in order to avoid the development of functional, aesthetic and even psychological disorders, with the aim of achieving therapeutic success and improving the quality of life of our patients [3,4].

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no potential conflicts of interest with respect to the authorship and/or publication of this article.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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