P30 A variant of the Seckel Syndrome: A case report

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Introduction

Seckel syndrome is an autosomal recessive disorder characterized by intrauterine growth retardation and post-natally by dwarfism, severe microcephaly, bird-headed profile with receding chin, prominent nose, joint defects, clubfoot, sparse hair, malformation of genitourinary tract and rectum, sweet disposition, mental retardation and hematological disorders. There is also a reduction in the number of blood cells. Dental anomalies of Seckel syndrome are crowded teeth with malocclusion, enamel hypoplasia and absence of some teeth.

Objective

A 16-year-old boy with Seckel syndrome visited our dental hospital for oral examination. His birth weight was 2.1kg. Microcephaly with bird face was present at birth. He had borderline mental retardation with normal motor development. He had hepatitis B, iron deficiency anemia with low hemoglobin values and hypospadia, but not the dwarfism was present (His height and weight were 157cm and 33.5kg). This could be an incomplete form or a variant of the syndrome. Intraoral findings were severe deep overbite, unilateral lingual crossbite and crowded teeth.

Case operation

Clinical oral examination and radiographs were obtained: craniofacial profile for analysis the craniofacial morphology, a panoramic radiograph for analysis the dentition. Impressions of upper and lower teeth were taken to make diagnostic models. Extra and intra oral photo were obtained. Fluoride vanish was applied for prevention dental caries.

Summary

A 16-year-old boy with Seckel syndrome was examined. His tooth Development and maturity progressed normally, but malocclusion was observed. Bleeding complication in invasive dental treatment can be occurred due to his low hemoglobin values. Consultation with the attending physician will provide characteristics of the disease and best approach to the treatment. The dentist should counsel patients and their parents to aware of the importance of oral hygiene in order to avoid the need for invasive dental care and reduce the number of visits to the dentist.