Comprehensive dental management in a Hallermann–Streiff syndrome patient with unusual radiographic appearance of teeth

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Abstract

Hallermann–Streiff syndrome (HSS) is a genetic disorder characterized by proportionate dwarfism, birdlike facies, hypotrichosis, skin atrophy, dyscephaly, bilateral microphthalmia, congenital cataracts, a narrow, weak, beaked nose, a hypoplastic mandible, and orodental anomalies. Occurrence is sporadic and distinct patterns of inheritance have not been found. This case report describes the dental management of a 3-year-old girl patient with HSS, who had unusual radiographic appearance of teeth. Furthermore, dental treatments and a 30-month follow-up period of the patient with this rare tooth structure malformation have been presented.

Key words: Dental anomalies, Hallermann–Streiff syndrome, orofacial characteristics

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Introduction

Hallermann–Streiff syndrome (HSS; OMIM 234100) is a genetic disorder characterized by proportionate dwarfism, bird-like facies, hypotrichosis, skin atrophy, dyscephaly (scaphocephaly or brachycephaly with frontal bossing), bilateral microphthalmia, congenital cataracts, a narrow, weak, beaked nose, a hypoplastic mandible, and orodental anomalies.[1] HSS was first described by Aubry in 1893, Hallermann in 1948, and Streiff in 1950. This syndrome is also called oculomandibulofacial syndrome, François syndrome, oculomandibulodyscephaly with hypotrichosis, Aubry syndrome I, and Ullric–Fremery–Dohna syndrome.[1-3]

Currently, the etiology of HSS is incompletely elucidated. Sclaroff and Eppley[1] suggested that it was a developmental disorder in the 5th-6th gestational week, correlated to either the first or second branchial arch syndromes, which affect ectodermal- and mesenchymal-derived structures. The incidence of the syndrome has been reported by Higurashi *et al*.[4] to be only one case in 27,472 newborn infants. Almost all cases are sporadic, and consequently, distinct patterns of inheritance have not been found.[5] Both sexes were affected equally.[3]

There have been more than 180 cases of HSS reported throughout the literature.[6,7] However, very few have addressed the orodentofacial features of the syndrome.

This case report describes the unusual radiographic appearance of teeth detected in both dentitions in a 3-year-old girl with HSS. Furthermore, the craniodentofacial findings, dental treatments rendered, and status after a 30-month follow-up period of the patient with this rare tooth structure malformation have been presented. The differential diagnosis of the syndrome and various treatment approaches are also discussed.

Case Report

A 3-year-old girl with a diagnosis of HSS by the Akdeniz University, Faculty of Medicine Department of Pediatric

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Genetics, was referred to our clinic for a mastication disorder, pain, and dental bleeding since 2 months.

She was born at the 38th week of gestation by vaginal delivery, the third child of healthy nonsconsanguineous parents. Her birth weight was 2300 g, birth height was 48 cm, and head circumference was 35 cm (25-50th centile). Her prenatal ultrasound showed micrognathia and retrognathia by the second trimester. No amniocentesis was performed. The medical history of her family was normal.

On physical examination, her weight was 11.6 g (<3rd percentile), and height was 84 cm (<3rd percentile). Proportionate short stature, brachycephaly, frontal bossing, microphthalmia, hypotelorism, strabismus, downs landing palpebral fissures, blue sclera, low set ears, long philtrum, small mouth, speech disorder, broad cheeks, hypoplastic nipples, left thumb shortening, and bilateral syndactyly of the 4th and 5th fingers were observed. Her face was characterized by a thin, sharp, and hooked nose, with microretrognathia, resulting in a characteristic birdlike appearance [Figure 1]. Furthermore, atrophic skin and sparse hair and eyebrows were noted.

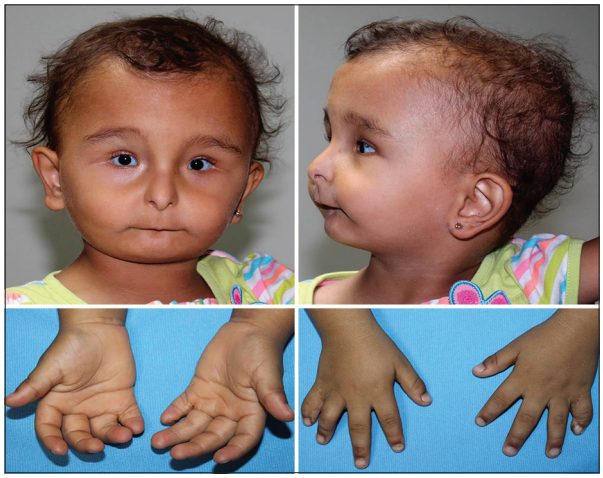

Figure 1: Bilateral syndactyly of the 4th and 5th fingers and facial view of the patient

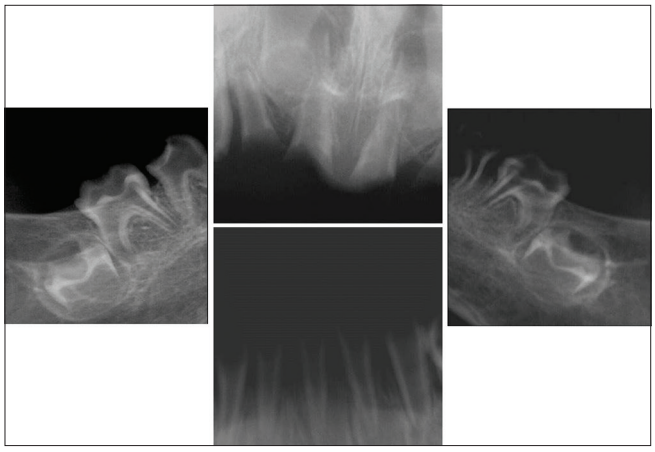

Figure 3: Radiographic examination revealed developmental malformations in the hard structures of teeth, causing a "ghost teeth" appearance

Her cranial radiograph revealed thin cranial bones, wormian bones, and microretrognathy. Her echocardiography detected a secundum atrial septal defect. Laryngomalacia, recurrent respiratory distress, and sleep apnea were also described in our patient's medical history. Both her abdominal ultrasonography and ophthalmologic examination were normal.

Her complete blood count and routine biochemistry tests for renal, liver, and thyroid function were within normal limits. Chromosome analysis of peripheral leukocytes using a high-resolution binding technique showed a normal 46, XX karyotype.

On intraoral examination, hypoplasias and attritions connected with hypoplasias were discovered in all of the deciduous teeth. In addition, caries in varying stages, most of them involving the pulp, were detected. Class II malocclusion and a high, narrow palate were revealed during the dental arch relation [Figure 2].

Radiographic examination revealed developmental malformations in the hard structures of the deciduous and permanent teeth (especially in dentin), causing a "ghost teeth" appearance [Figure 3].

Due to the good psychomotor development of the patient, she was cooperative during her first visit to our dental clinic, although she had a speech disorder. The IQ assessment of the patient was evaluated, and normal values were found.

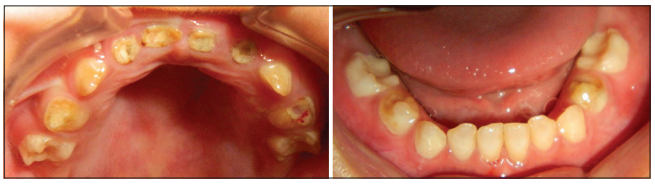

Figure 2: Intraoral appearance from the first visit of the patient

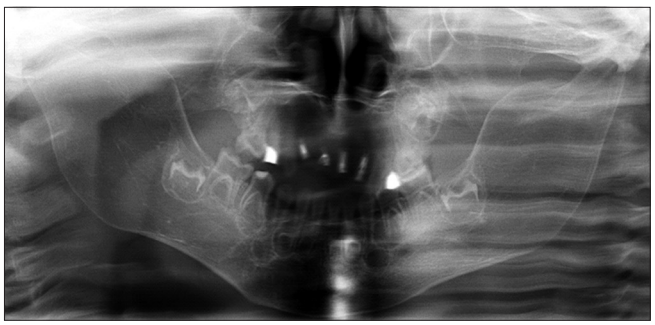

Figure 4: Orthopantomographic view of the patient at the beginning of the dental treatment

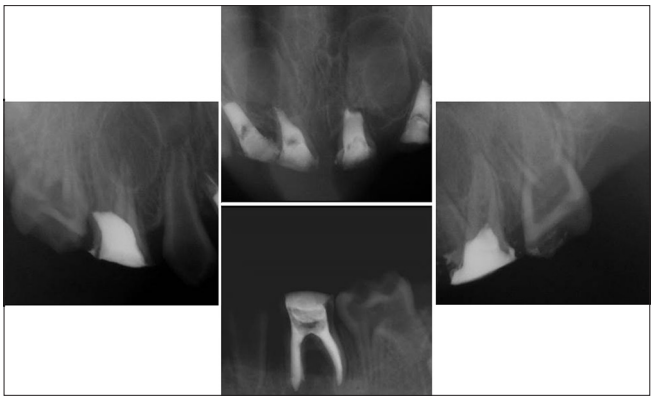

Figure 5: Periapical radiographies of the teeth after dental treatments

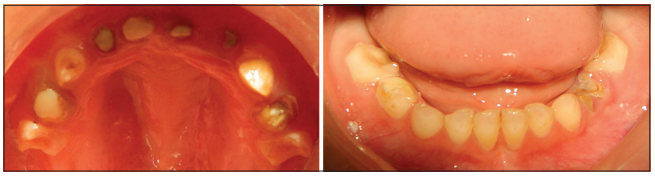

Figure 6: Intraoral appearance of the patient after 1 year for control visit

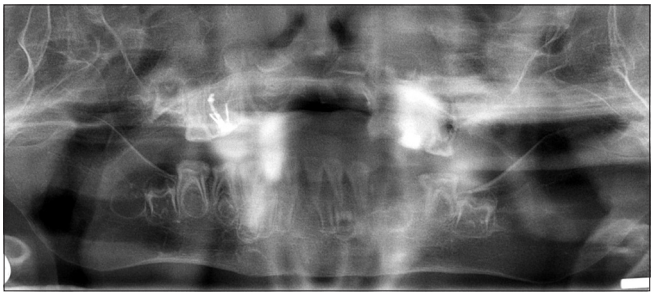

Figure 7: Orthopantomographic view of the patient after a 30 months follow-up

Thereafter, a dental treatment plan was established, and oral hygiene instructions and dietary recommendations were presented to her parents.

Restorative treatment of primary right mandibular first molar (84), endodontic treatments primary maxillary right and left central and lateral incisors (51, 52, 61, 62) and primary mandibular left first molar (74) and pulpotomy of primary maxillary right and left first molars (54, 64) were performed under clinical conditions [Figures 4 and 5]. Fluoride varnish was applied on the teeth during subsequent appointments; the patient was fully cooperative. Because of the known dental hypoplasia and developmental malformations of her teeth, exceptional care was taken not to damage any dental structures during treatment.

During the follow-up period, rapidly progressive root resorption was detected in endodontically-treated primary mandibular left first molar (74) and hence that at the end of 1 year, only some root fragments remained. The following year, an infection was found in primary maxillary right first molar (54) and root canal treatment was performed. Furthermore, progressive root resorption was detected in the primary maxillary anterior incisors (51, 52, 61, 62) that treated endodontically [Figures 6 and 7].

The patient, who is now 5 years old, is still followed-up for care of her deciduous teeth and guidance for healthy eruption of her permanent teeth.

Discussion

François[8,9] listed seven positive and five negative findings to note for a diagnosis of HSS after reviewing 22 patients. Positive signs are dyscephaly and bird face appearance, dental abnormalities, hypotrichosis, atrophy of the skin localized to the head and nose, bilateral microphthalmia, bilateral congenital cataracts, and proportionate dwarfism. In addition, it was reported that patients diagnosed with HSS also demonstrate facial dysmorphism, low birth weight, and growth retardation. Our patient showed all the characteristic findings except bilateral congenital cataracts. Five negative signs were also described by François[8,9] as differential diagnostic criteria for HSS. These include the absence of (I) auricular anomalies, (II) palpebral anomalies, (III) premature arteriosclerosis, arthrosis, deformities of joints, muscular atrophy, (IV) nail and extremity anomalies, and (V) mental retardation. No negative signs were seen in our patient.

The differential diagnosis of HSS from progeria and progeroid syndromes, mandibulofacial dysostosis, and pseudoprogeria is as follows. Progeria differs from HSS by having premature atherosclerosis, nail dystrophy, chronic deforming arthritis, acromicria, and normal ocular findings. Mandibulofacial dysostosis usually has ear anomalies and lower eyelid colobomas. Pseudoprogeria/HSS syndrome with an autosomal recessive trait shares similar findings, but it has a normal appearance at birth, psychomotor delay, and severe spastic quadriplegia.[7]

Orodental anomalies reported in the literature (50-85% of the cases) are microstomia, a small and retracted tongue, mandibular hypoplasia, a high arched palate, class II malocclusion, open bite, hypoplasia of deciduous and permanent teeth, absence of teeth, persistence of deciduous teeth, supernumerary teeth, natal teeth, malformed teeth, and severe and premature caries.[2,3,9-11] In our case, microstomia, a small and retracted tongue, mandibular hypoplasia, a high arched palate, class II malocclusion, open bite, hypoplasia of deciduous and permanent teeth, malformed teeth, and severe and premature caries were seen.

The hard tissues of the teeth were determined to be developmentally malformed, causing all deciduous and permanent teeth to have a radiological appearance of "ghost teeth." Until now, only one similar case had been reported. A 15-year-old girl had decreased thickness of enamel and dentin, markedly enlarged pulp chambers and stunted root canals, creating a "ghost teeth" view.[12] In addition, intraoral examination revealed hypoplasic teeth, similar to our case. There should be more case reports to determine the characteristic features of HSS.

In our patient, dental treatments were performed under local anesthesia because, in this syndrome, the greatest anesthetic challenge lies in the maintenance of the airway. Narrow upper airway associated with the craniofacial configuration, narrowing of the posterior oropharynx, and secondary glossoptosis can lead to complete airway obstruction. In addition, microstomia and an undeveloped temporomandibular joint can make laryngoscopy difficult. Severe respiratory embarrassment and obstructive sleep apnea were also noted.[6,8]

Examination of the few publications available in the related dental literature shows that orthodontic/orthognathic, surgical, prosthetic (implants, prostheses, etc.), and conservative approaches are used to treat the patients' dental problems.[13-15] Each of these therapeutic approaches has its own advantages and disadvantages. Conservative treatment should initially be given preference, considering the multiple systemic abnormalities. Conventional treatments under local anesthesia, as performed in our patient, should be possible in several HSS patients. In the presence of various dental malformations and the high risk of caries, personalized oral health plans, and regular routine follow-up starting at the time of HSS diagnosis are of particular importance.

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