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Hallermann-Streiff Syndrome: A case report from Turkey

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Abstract
Hallermann-Streiff Syndrome (HSS) is a rare disorder characterized primarily by head and face abnormalities, with dental abnormalities also present in 50-80 percent of cases. The first description seems to have been made by Aubry in 1893. HSS was first described completely in 1948 by Hallermann, and then in 1950 by Streiff. We report a 4-year-old girl with HSS, presenting oro-dental characteristics and describe the treatment modalities. An interdisciplinary approach including early preventive-care programmes, detailed oral hygiene instructions, dietary recommendations, counselling of the parents and regular dental visits are the essential procedures.

Key words: *Hallermann-Streiff Syndrome, oro-dental characteristics, preventive-care program.*

Introduction
The first description for Hallermann–Streiff Syndrome (HSS) seems to have been made by Aubry in 1893. HSS was first described completely in 1948 by Hallermann and then in 1950 by Streiff (1).
Seven essential signs described by Francois as diagnostic criteria for HSS are presented in (table 1) (2).
In the course of neonatal period and infancy, death may occur due to respiratory challenges which arise in consequence of small nares and glossoptosis secondary to micrognathia and tracheomalcia in patients with HSS (3).
HSS is a rare disorder that is characterized primarily by head and face abnormalities. Patients show bird-like facies, dental abnormalities and hypotrichosis with various ophthalmic abnormalities (2).
Dental abnormalities are seen in 50-80 percent of the cases and include supernumerary and neonatal teeth, premature eruption of primary dentition, agenesis of permanent teeth and enamel hypoplasia. Malocclusion, crowding, anterior displacement or absence of condyles, mandibular hypoplasia, high arched palate and small mouth are the other remarkable oro-dental characteristics (4,5).

In the literature, there are limited reports concerning oro-dental implications, the most favorable treatment procedures and future perspectives for HSS. In this paper, we report a 4-year-old girl with HSS and present oro-dental characteristics and express the treatment modalities of this unusual syndrome and thus provide additional data for the literature.

Table 1. Diagnostic criteria for HSS.

1	Dyscephalia and bird-like facies
2	Abnormal dentition
3	Hypotrichosis
4	Atrophy of skin especially on the nose
5	Congenital cataracts
6	Bilateral microphthalmia
7	Proportionate dwarfism

Case Report

A 4-year-old girl with congenital HSS was referred to the Department of Pediatric Dentistry, Faculty of Dentistry, Süleyman Demirel University because of her dental treatment necessity. This baby was the first child of consanguineous parents (cousins from second rank) and was born by normal spontaneous delivery following 28th-29th weeks of eventful pregnancy. In first trimester; the mother had ampicillin-sulbactam treatment twice because of tooth infection. The birth weight and height of the baby were 1.6 kg and 44 cm, respectively and her head circumference was not known. The mother was 26-year-old and healthy however, the father was 31-year-old with a medium loss of hearing due to meningitis. There was no similar phenomenon in the family.

Her medical history revealed that a sistolic murmur was present on cardiovascular system and her chromosome analyses were normal. The mother stated that, wheezing and respiratory embarrassment had begun when the baby was two-month-old and have increased at the nighttime. The history of sleep apnea and recurrent lower respiratory system infection were recorded.

Her physical growth and development was retarded however, her mental development was normal. There were spoon nails and clubbing and she had simian crease on her left palm. The sparse hair on head, eyebrows and eyelashes was visible. The skull was brachycephalic with frontal bossing. Skin of the face appeared atrophic. There were hypertelorism, blue sclera and hypotrichosis of the scalp, brows and lashes. Her eyes were normal without microphthalmos and congenital cataract. The nose was thin and beak-shaped (Fig.1).

A retrognathia and small mouth opening was present and she had a hypoplastic mandible. There was a cleft on the symphysis region of the mandible. When she was two-month-old, the eruption of the first teeth was observed on the mandibular incisors region. Pulpy feeding diet was observed and brushing period was not systematic. Examination of the oral cavity demonstrated the following findings; high arched narrow palate, mobility of the right-left mandibular segments and crowding of the mandibular anterior teeth due to the absence of fusion on the symphysis region, anterior open-bite, expanded severe caries and plaque accumulation on all teeth (Fig.2 and 3).

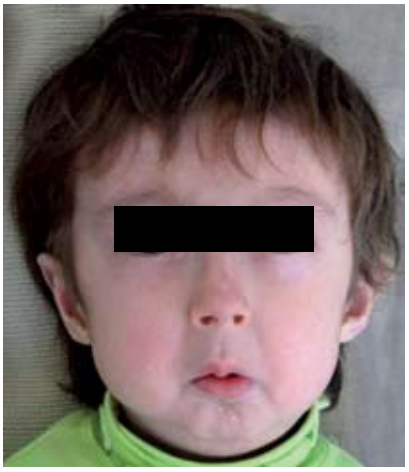

Fig. 1. Facial view of the patient.

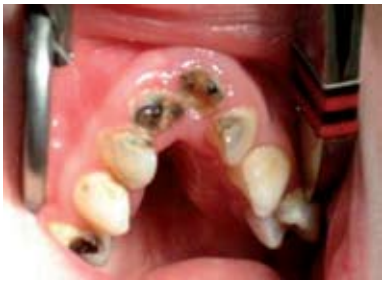

Fig. 2. The view of oral cavity (the maxillary segment).

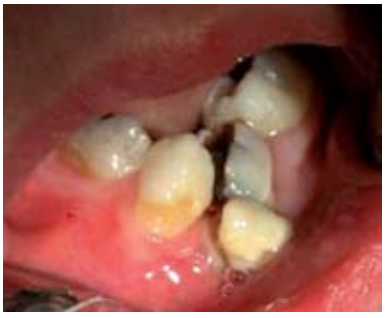

Fig. 3. The view of oral cavity (the mandibular segment).

Cooperation problems, respiratuar challenges, small mouth and mobility of the mandibular segments due to the absence of fusion on the symphysis region affected all treatment procedures and radiographs therefore, we could not take radiographs and intra-oral photographies adequately and could duplicate her teeth in three portions.

She was immediately included in a preventive-care programme. Detailed oral hygiene instructions and dietary recommendations were provided at the first appointment counselling to her mother or father. She and her parents could not come to our clinic regularly because they resided in a village far away from Isparta and she had frequently respiratuar problems and was taken to the hospital.

Restorative and preventive procedures were performed on following appointments. Tooth 51,61,74 and 84 with severe caries, excessive substance loss and infection were decided to be extracted. On the maxillar segment; tooth 54,52 and 62 with dentine caries were restored with compomer (Dyract Extra, Dentsply, Konstanz, Germany) and tooth 64 with dentine caries was restored with silver-reinforced glass ionomer cement (Argion, Voco, Cuxhaven, Germany).

On the mandibular segment, limited treatment was fulfilled because of the mobility of the right-left mandibular segments and the crowding of the mandibular anterior teeth due to the absence of fusion on the symphysis region. Tooth 75 and 85 had severe caries and tooth 85 was restored temporarily with glass ionomer cement (Ketac-cem, 3M Espe, Seefeld, Germany) but tooth 75 could not be treated. Fluoride varnish (Bifluoride 12, Voco, Cuxhaven, Germany) was applied to the remaining teeth and a follow-up programme was planned for her. Follow-up views at 2 years revealed the durable and sound restorations and the patient remained asymptomatic.

Discussion

Seven essential signs were described by Francois as diagnostic criteria for HSS (2). Our patient had five of the seven signs of this syndrome and there were not ocular abnormalities except for hypertelorism and blue sclera. Mental development is usually normal, but exceptions are not uncommon (4). Physical growth and development was retarded however, mental development was normal in our patient.

An asymmetric second branchial arch defect which appears in the time of fifth or sixth gestational week seems to be effective in the aetiology of HSS. The inheritance model is unknown and this syndrome seems to be a sporadic mutation. Familial cases have been reported. However, currently avaible data are considered more probably suggesting an autosomal recessive inheritance (6). Consanguineous parents (cousins from second rank) and the mother's ampicillin-sulbactam treatment twice because of tooth infection in first trimester might have taken a part in this case.

There are persistent active oral pathogens because the patients with HSS are prone to repeated respiratory tract infections and early deaths due to respiratory infections have been reported and pre and post-operative antibiotic cover is recommended (1). Early death of the patients with HSS due to respiratory challenges necessitates the improvement of life quality.

In this syndrome, the reform of deformities which complicates the life is considered first. There are HSS Support Groups and recommendations for respiratory challenges, general anesthesia, ocular abnormalities and rhinoplasty are generally presented however, data on dental procedures, treatment alternatives for the mandibular deformities as in our patient and future perspectives are not satisfactory. Very few cases have been described in the dental literature and surgical and prosthetic interventions are scheduled at completion of growth to solve the skeletal discrepancy and for occlusal rehabilitation (7). Unsatisfactory data for HSS in the literature together with cooperation problems, respiratuar challenges, small mouth and mobility of the mandibular segments due to the absence of fusion on the symphysis region affected all treatment procedures and we fulfilled limited treatment.

The predisposition to severe caries, together with other problems, makes it imperative that young patients must be included in a strong prevention program as early as possible. An interdisciplinary approach, early preventive-care programs, detailed oral hygiene instructions and dietary recommendations counselling to the parents and regular dental visits are the essential procedures for the patients with HSS.

The authors who treated patients with unusual syndromes such as HSS should share and present their treatment approaches and applications with other authors.

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