Hallerman Streiff Syndrome : A Rare Clinical Case Report

Abstract

Hallermann Strieff Syndrome, also known as Occulo-mandibulo Dyscephaly syndrome is a rare clinical entity of unknown etiology which involves multiple congenital abnormalities affecting chiefly the head and face, with over 150 cases reported in the literature worldwide. Dyscephaly, micro-pthalmia, cataracts, hypotrichosis, cutaneous atrophy, beak shaped nose and a typical bird face are the main features of the syndrome. We report a case of 13year old girl with Hallermann Strieff syndrome, with special considerations on oro-dental findings. Clinical, radiographic and cephalometric analysis revealed partial anodontia, irregularly developed teeth with altered morphology of the crowns, high arched V- shaped palate, micrognathia and hypoplasia of the mandible. The purpose of this case report is to discuss the features of Hallermann Strieff syndrome with the differential diagnosis and the overall management of the patient.

Key Words

Occlusomandibulodyscephaly, micro-pthalmia, anodontia, micrognathia.

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Introduction

Hallermann Streiff Syndrome is a rare congenital disorder characterized primarily by head and face abnormalities, and dental abnormalities present in 50 to 80% of the cases.[1] The first published record seems to have been made by Aubry[2] in 1893, but was described completely in 1948 by Hallerman[3] and then in 1950 by strieff[4], who after separating the syndrome from progeria and mandibulofacial dysostosis coined the term Hallermann Streiff Syndrome (HSS)[5]. Over 150 cases have been reported in the medical literature with relative no sex predilection. In 1958, Francois[6] reviewed the literature, analyzed the manifestations of 22 published cases and described diagnostic criteria for this syndrome which include

1. dyscephalia and bird like facies.
2. abnormal dentition
3. hypotrichosis
4. atrophy of the skin especially, on the nose.
5. congenital cataracts
6. bilateral micropthalmia
7. proportionate dwarfism

Well documented cases of HSS have all been sporadic[7] not associated with chromosomal anamolies. Nothing much was known about the cause. It seemed that this syndrome did not have typical autosomal dominant or recessive trait, despite its sporadic manifestations[6]. The evidence implicating contributing factors in HSS is questionable. There were familial case reports, but all were either atypical cases or poorly documented[9].

In the literature there are limited reports concerning orodental implications, the most favorable treatment procedures and future perspectives for HSS[8]. In this paper we report a 15 year old girl with HSS and present the orodental characteristics with treatment modalities of this unusual syndrome and provide additional data for the literature.

Case Report

The case involved a 15 year old female, who reported with the chief complaint of missing teeth and pain in the lower right quadrant. Patient did not give history of any past dental treatment. The birth history showed a normal vaginal delivery after an uncom-plicated full term pregnancy of a non consanguineous marriage. Her mother's history revealed no record of any systemic or drug administration during her pregnancy. Her parents and younger sister were normal with no specific findings.

Medical history revealed feeding problems during infancy and symptomatic treatment taken for recurrent respiratory infections. The patient has not achieved menarche till now. She complains of decreased visual aucity for which no treatment was taken till date. Her physical as well as mental growth was retarded.

Extraoral Examination

Revealed frontal bossing, small and thin face slanting palpaberal fissures, bilateral micropthalmia, beak shaped nose accom-panied with microstomia and retro-gnathia. The skin of the face appeared dry and thin. There was slight bluish tinge visible on the sclera and hypotrichosis of the scalp, brows and lashes were apparent.

Intraoral Examination

Revealed retrognathia and a small mouth opening, a high arched V shaped palate. Partial anodontia was present, with remaining teeth exhibiting enamel hypoplasia, yellowish discoloration and were irregularly developed with altered morphology of the crowns thereby making their identification difficult.

Orthopantomography

Revealed absence of numerous permanent teeth and a retained deciduous tooth, with no permanent tooth buds. All present teeth showed features of ghost teeth, decreased enamel and dentine thickness, marked enlarged pulp chambers and canals, stunted roots. Generalized horizontal bone loss was present in both the arches. Mandibular body was small with no antegonial notch. The mandibular rami were hypoplastic and lateral ceephalogram showed for calvarial thickening, midfacial hypoplasia and frontal bossing.

To confirm the diagnosis the patient was referred for gynecological, opthalmological and ENT opinion. Diagnosis of Hallerman strieff syndrome was confirmed and treatment planning done. First and foremost, every effort was made to preserve the teeth present and prevent any unfavorable sequence. Detailed oral hygiene instructions and dietary recommendations were provided at the first appointment to her mother. Oral prophylaxis was performed. Teeth with severe caries, excessive substance loss and a retained deciduous were extracted.

For the replacement of missing teeth, the major challenge was poorly formed teeth with very less crown height and substance. Surgical and prosthetic interventions were scheduled at completion of growth to solve the skeletal discrepancy and reconstructive procedures can be done after the adolescent growth period is complete. Regular follow ups will be done for the patient and the option of overdenture and implants could be considered after the growth is completed. For the time being an interim partial denture was fabricated for the patient to improve esthetics and function.

Discussion

Seven essential signs were described by Francois6 as diagnostic criteria for HSS. Our case had six of them which includes Dyscephelia and bird face, dental anamolies, proportionate nanism, hypotrichosis, atrophy of the skin, bilateral micropthalmia with no congenital cataracts. Additionally our case showed mental retardation (IQ less than 61/65) which is reported only in 15% of the cases[7,8].

Differential diagnosis include Progeria and other progeriod syndromes such as DeBarsy, Hutchinson Guilford, pseudoprogerial hallermann streiff syndrome(PHS), Cockaynes syndrome as well as mandi-bulofacial dysostosis, cleidocranial dysos-tososis and pyknodysostosis[7].

The PHS syndrome, in addition has severe spastic quadriplegia and appearance at birth is normal except for eyebrows and eyelashes.

Progeria differs from hallermann streif syndrome because of premature arteriosc-lerosis, nail dystrophy, acromicria, chronic deforming arthritis normal eyes. Mandi-bulofacial dysostosis differs from HSS as it usually has lower eyelid colobomas and associated ear anamolies.

Dental anamolies are common and may include natal and neonatal teeth, absence of teeth, supernumerary teeth, malformed teeth and enamel hypoplasia. Severe agenesis of permanent teeth and delayed eruption of existing teeth are frequent findings[10]. Hence young patients must be

included in a strong preventive programme as early as possible. An interdisciplinary approach, early preventive programme, detail oral hygiene instructions and dietary recommendation counseling to the parents with regular follow ups are essential procedures for patients with HSS[11].

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 and ocular abnormalities and rhinoplasty are generally presented however data on dental procedures, treatment alternatives for the mandibular deformities and future perspectives are not satisfactory[1].

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. Unsatisfactory data for HSS in the literature together with cooperation problems, respiratory challenges, small mouth opening affected all treatment procedures[12].

There is no cure reported for HSS, but one should pay special attention to opthal-mological and ENT examination should be performed for the early diagnosis for eye and respiratory complications, as with other genetic syndromes, genetic counseling should be performed in all affected patients.13 Clinical management must focus on more life threatening and developmental issues earlier on, and reconstructive procedures can be done after the adolescent growth period is complete.[14]

The authors who have treated patients with unusual syndromes such as HSS should share and present their treatment approaches and applications so that such patients may benefit from them.

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Legends

Fig 1. Facial Manifestations of the patient, frontal view

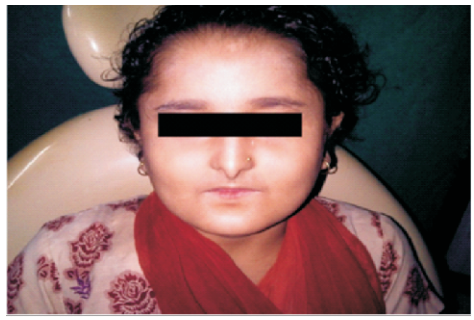

Fig 2. Lateral view

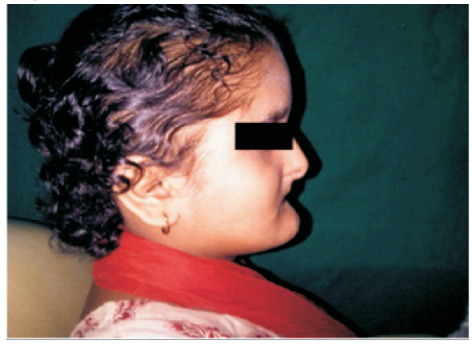

Fig 3. Intraoral view (maxillary pre operative)

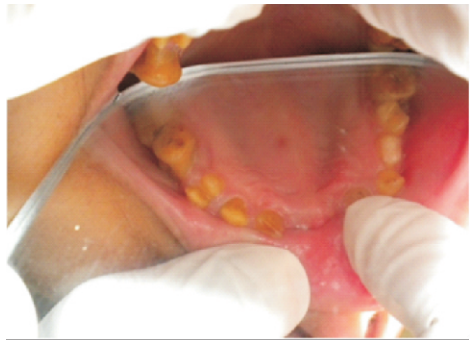

Fig4.Intraoral view (mandibular pre operative)

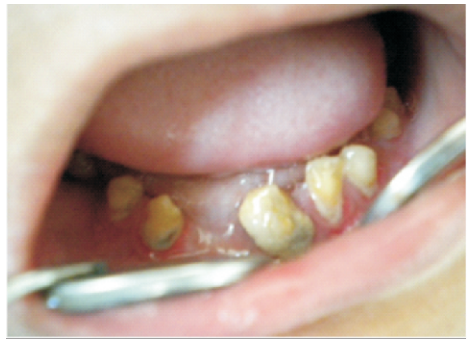

Fig 5. Lateral cephalogram

Fig 6. Gum strippers

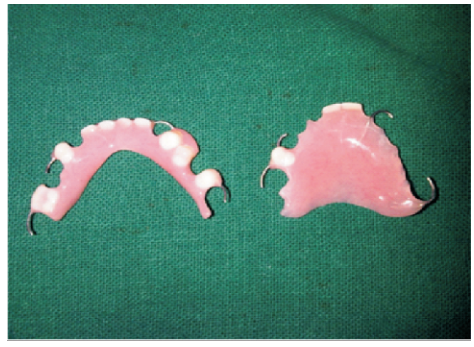

Fig7. Intraoral view (maxillary post operative)

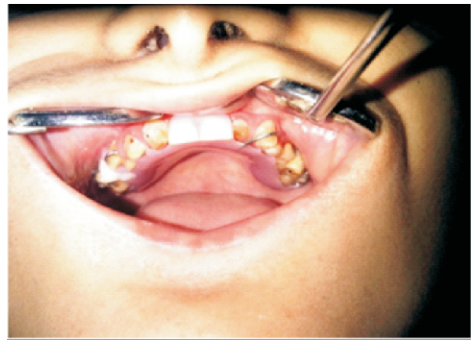

Fig8.Intraoral view (mandibular post operative)