Atypical Hallerman-Streif Syndrome: A Case Report

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Hallerman Streif syndrome is a rare congenital disorder characterized by dyscephaly, dental anomalies, proportionate nazism ,hypotrichosis, cutaneous atrophy limited to the head ,bilateral congenital cataracts and bilateral microphthalmia .Despite the marked craniofacial characteristics and oral findings,a relative lack of reports in the dental literature has been noted. In this article, a case of a 8 year old boy with dental problems is described.
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INTRODUCTION

Hallerman – Streif syndrome also known as oculo mandibulo dyscephaly is a rare congenital disorder characterized by dyscephaly, hypotrichosis, microphthalmia, dental anomalies, cutaneous atrophy restricted mainly to head and neck and most obvious over the nose and bilateral congenital cataracts. Aubry earlier described the condition in 1893.[1] Hallerman in 1948[2] and Streif in 1950[3] separated the syndrome from progeria and mandibulofacial dysostosis

INCIDENCE

Over 150 cases have been reported in the medical literature so far with relatively no sex predilection.

LITERATURE REVIEW

A genetic influence expressed with all autosomal dominant pattern of inheritance has been suggested by Spoerl[4] and Koliopoulos.[5] While familial as well as parental consanguinity incidences of this syndrome have been described by François.[6]

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A mal development of the first branchial arch during the 5th to 7th weeks of gestation has been suggested as being responsible for the pathogenesis since there are changes corresponding to both mesenchymal and ectodermal structures in origin. A discrepancy in the pair of D group chromosomes has been reported by Carones, however, no other anomalies in the chromosomal material have been found.[7]

Although the pathogenesis of oculo mandibulo dyscephaly(OMD) is far from elucidated, a sporadic mutation is thought to be responsible for the numerous anomalies of face, skull and skin. Probably the genetic defect also underlies the OMD associated dental abnormalities. The early loss of dental lamina can be considered an additional expression of a general ectodermal hypoplasia, hypotrichosis and skin atrophy being other expressions of this OMD associated anomaly.

GENERAL FEATURES

Face: patients present with a characteristic facial appearance with a thin beak like nose and may have a tendency to septal deviation. Receding chin is a characteristic finding which is a result of the mandibular hypoplasia.[8] This hypoplasia involves both body and ramus of mandible while the condyles are anteriorly placed and occasionally absent.[9]

Cranium: Developmental anomalies that involve craniofacial structure give genesis to a marked dyscephalia characterized by a large cranial vault and a small facial skeleton.[10]

Radiographic screening of young children with this syndrome reveals a poorly ossified skull, while sutural closures varies considerably. Early closure of the sagital suture is responsible for a long, narrow head resulting in scaphocephaly, with a bulging occiput and forehead. Delayed closures of fontanelles with persistently wide sutures give genesis to brachycephaly with frontal boss-

ing and parietal prominence, which is frequently found characteristic of this syndrome. Measurement of head circumference is usually normal or slightly smaller.[10]

Eyes: Ocular findings include microphthalmia and congenital cataracts in the majority of cases.[5, 11] Other ocular signs encountered with less frequency are the presence of nystagmus, strabismus and glaucoma.[11, 12,13]

Skin: Ectodermal manifestations associated with this syndrome include hypotrichosis and cutaneous atrophy. The scalp hair, eyelashes and eyebrows are sparse with alopecia being most prominent in the frontal and occipital areas of skull.[13] The skin is thin and atrophic, particularly in the scalp and over the nose.[6]

GROWTH AND DEVELOPMENT

A proportionate decrease of growth is noted for both males and females resulting in a short stature. The final height of females has a mean of 152.4 cm with males being 2.5-5.0cm taller.[14] (Fig.1)

OTHER FEATURES

Genital organ anomalies have been shown including hypogenitalism, cryptorchidism, clitoral enlargement and breast atrophy.[15] In 15-31% of the cases even mental retardation has been described.[6,11]

Other anomalies have also been reported such as: Skeletal defects (10%-15%), cardiac defects (2%–9%), hematopoietic abnormalities (7%), pulmonary anomalies (3%).

INTRAORAL FINDINGS

- The palate is high and narrow
- Hypoplastic mandible with extremely obtuse mandibular angle.[10]
- The abnormal position of the condyles in conjunction with the anatomical changes in the temporal bone and glenoid fossa may result in limited mouth opening.
- Oral cavity is small due to the midface deficiency and mandiblular hypoplasia.
- Developmental anomalies that affect the number, the shape, the structure and position of the teeth is a consistent findings in patients with Hallerman Streif syndrome.

The dental anomalies can be divided into primary and secondary events.

Primary: The agenesis of almost all permanent teeth can be explained by the early disintegration of the dental lamina. This lack of permanent teeth can explain the persistence of primary teeth (François 1958, Fall & Schall 1960, Caspersen & Warburg 1968).

Secondary: The premature loss of Hertwig's root sheath coupled with an irregular architecture of the root dentin is a secondary feature. The premature eruption in which the teeth become exposed to the oral cavity in a very immature and only partly calcified stage are thus, prone to deformation, tooth malformation hypoplasia and impaction.

Figure 1. Photograph of the patient with his sibling who is five years old.

CASE REPORT

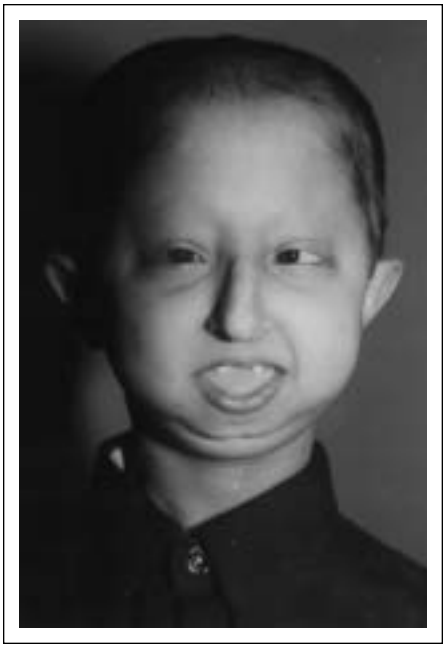
An 8 year old male patient who reported to the department of Pedodontics and preventive dentistry, A. B. Shetty Memorial Institute of Dental Sciences Mangalore with a chief complaint of pain in relation to the lower anterior teeth. The child was born from a full term normal pregnancy and had an uncomplicated delivery. No consanguinity was reported and the family history was negative. He was the first-born child of the family.

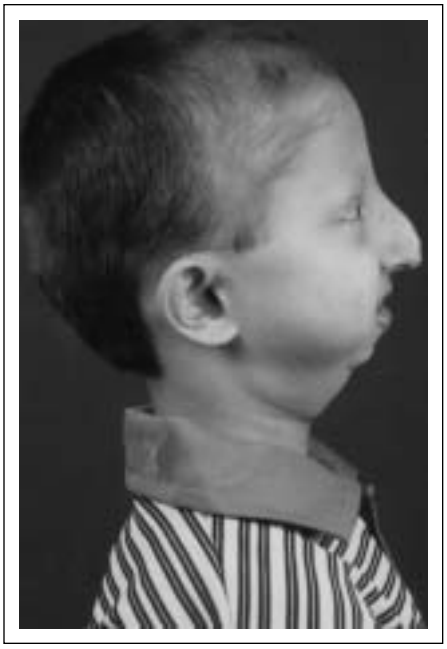
Nothing relevant was revealed in medical history except for ophthalmic consultation for nystagmus. Patient has been on correcting glasses for 4 years and is under regular ophthalmic care.

On extra oral examination he was found to present, dyscephaly (Odd shaped bulging of the skull), bossing of parietal bones, hypotrichosis of scalp, alopecia in frontal and occipital areas, absence of the eyebrows, bird shaped facial appearance with marked mandibular hypoplasia and thin, sharp nose deviated to left side, thin lips, microphthalmia and double cutaneous chin (Figures 2 and 3).

ON INTRAORAL EXAMINATION:

- Localized marginal gingivitis was present in relation to lower anterior region with fair oral hygiene.
- High vaulted constricted and 'V' shaped palate with no evidence of bony or mucosal clefting.





Figures 2 and 3. Photographs showing extra oral findings of the patient.

Dental history revealed the presence of 3 natal teeth (2 upper and one lower) which were extracted 15 days after birth.

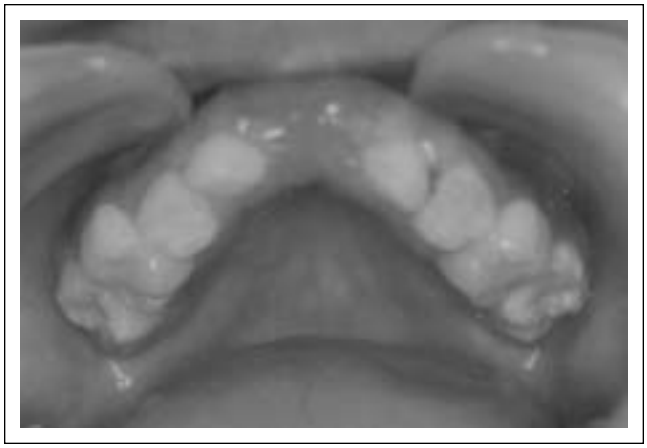
All the remaining primary teeth were present with the eruption of 1st permanent molars on all four quadrants.(figures 4 and 5). Dentition at the time of examination was free of caries and showed crowding of lower anterior teeth with lingual eruption of left central incisor and rotated canines.

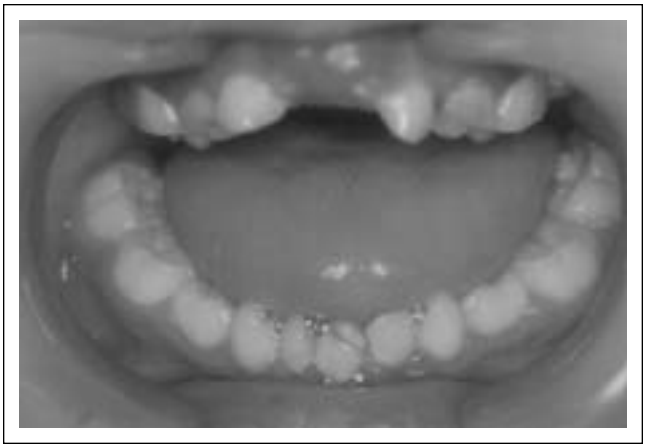
The panoramic X ray shows: Absence of succedaneous permanent teeth, presence of the teeth germs of upper right and left permanent second molars the presence of a tooth in between upper primary lateral incisors, condylar hypolasia (Figure 6).

Cephalometric tracing shows underdevelopment and retrusion of the entire maxillo-mandibular complex in both anteroposterior and vertical dimensions (Figure 7).

TREATMENT

Understanding the complex nature of this presentation, we decided to initially treat the dental problems only and keep the patient under constant observation for timely treatment wait for tooth eruption and observe the growth and development of the child. Since there is congenital absence of several permanent teeth, particular concentration in preventive measures was necessary to maintain the primary teeth free of caries





Figures 4 and 5. Photographs showing intra oral findings of the patient.

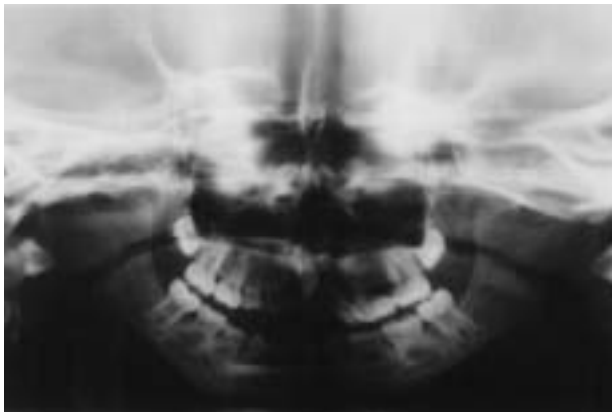


Figure 6. Panoramic X Ray of the patient showing absence of succedaneous permanent teeth.

and help the patient acquire good oral habits. The maxilomandibular hypoplasia present may definitely require orthognathic surgery after the child's growth is completed.

CONCLUSION

Management of the multiple dental and skeletal problems these patients present makes critical the follow up of their dental needs during childhood and adolescence.

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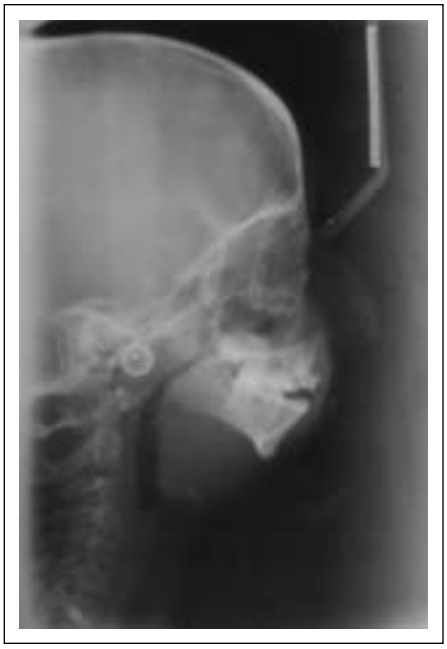

Figure 7. Photograph of lateral cephalogram of patient showing condylar hypoplasia.

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