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Case Report

CASE REPORT: HALLERMANN STREIFF SYNDROME

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ABSTRACT

The Hallermann Strieff syndrome (HSS) is a rare congenital disorder characterized by distinctive craniofacial malformations and significant orodental abnormalities. Very few such cases have been reported in the literature. Most cases of hallermann–strieff syndrome occur randomly, for unknown reason, may be the result of mutations, or changes in the genetic material. HSS is associated not only with developmental anomalies involving structures of ectodermal origin (face, skull, hair, skin, eyes and teeth) but also affects overall growth and development. One of such rare case of hallermann streiff syndrome reported at Department of Orthodontics, Government Dental College and Hospital, Ahmedabad with chief complaint of proclination of maxillary anterior is presented.

INTRODUCTION

Hallermann-Streiff syndrome (HSS),an uncommon anomaly featuring oculo-mandibulo-cranial malformation. HSS was first incompletely described by Aubry in 1893 as oculomandibulodyscephaly syndrome, later described by Hallermann in 1948 and streiff in 1950, the term "bird face" was used because of the peculiar facial appearance[1].HSS results from a developmental disturbance that arises between the 5th and 7th week of embryonic life and affects the cephalic ventral extremity at themoment when development of facial bones and of lenses is at the highest degree, thus involving bothectoderm and mesoderm[2]. Hallermann-Streiff syndrome is considered to be a congenital disease characterized by dyscephalia, dental anomalies, proportionate nanism, hypotrichosis, atrophy of the skin, bilateral microopthalmia, and congenital cataract.[3]since the major symptom in all patients with this syndrome is cataract, most cases have been reported in the ophthalmologyliterature. Radiographic findings of the syndrome include hypoplasia of both the mandible and the maxilla and dysrhaphism ofthe suture[4].

The dental literature with respect to the Hallermann- Streiff syndrome has reported two additional radiographic signs,1: anterior displacement of the temporomandibular joint (TMJ), first reported by van Baleri,[5] and2: close proximity of the root apices of the molar teeth to the lower border of the mandible, reported only by Hutchinson[6]. Polymorphism of clinical signs is typical of HSS. Therefore, even if it is a rare disease, it requires an exact diagnosis in order to evaluate prognosis and implications of a probable genetic transmission.[7, 8, 9]

In 1958, Francois gave the diagnostic criteria of this syndrome. Around 150 cases have been reported in the literature. A case of 15 year old boy with classical signs and symptoms of Hallermann streiff syndrome is presented.

Case Report

A 15 year old boy reported with the chief complaint of proclination of upper anterior teeth with no relevant dental history. As per the history, the patient was born after an uncomplicated full term pregnancy of a non-consanguineous marriage. His mother's obstetric history revealed no record of systemic disease or drug administration. His siblings were normal. His medical history revealed decreased vision and difficulty in breathing during sleep.

Physical examination of the patient's face revealed frontal bossing, small face, low set flared ears, small beaked nose, microopthalmia, opaciy of lens bilaterally, microstomia, retrognathia and convex profile . Skin of the face appears dry with hypotrichosis of scalp and eyelashes (fig 1). The patient has a propionate nanism. Intraoral examination revealedfull set of permanent teeth in maxillary arch and hypodontia of right central and lateral incisor and left lateral incisor in mandibular arch, scissor bite in right premolar region, unilateral chewing habit on left, fluoresced cervical margins of incisors, white

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patches on left buccal mucosa, large tongue with tongue tie, and small mandible.(fig 2)

Orthopantogram shows absence of lower right central incisor and both mandibular lateral incisors and impacted third molars in all quadrants. (fig 3 a). Lateral cephalogram reveals retrognathic, micro gnathic maxilla and mandible with skeletal class I relationship, vertical growth pattern, increased overjet and overbite. Upper incisors proclined and forwardly placed bodily, lower incisors retroclined (fig 3 b). Hand wrist radiograph indicates SMI stage III (Skeletal age estimate to be 12 ± 1years and chronological age 15 years). The radiographs of long bones and chest showed no significant findings. Chromosome analysis showed no evidence of structural or numerical abnormality in any of the chromosomes, except prominent satellite on chromosome 22, which is considered to be a normal variant. Thus a diagnosis of Hallermann streiff syndrome was made.

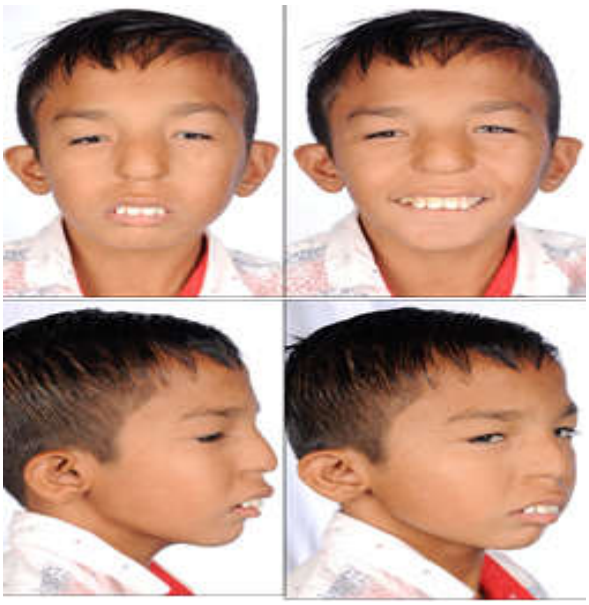

Figure 1 Facial photographs

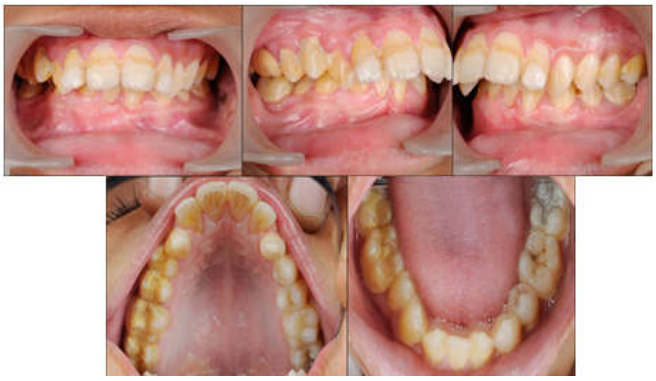

Figure 2 Intraoral photographs

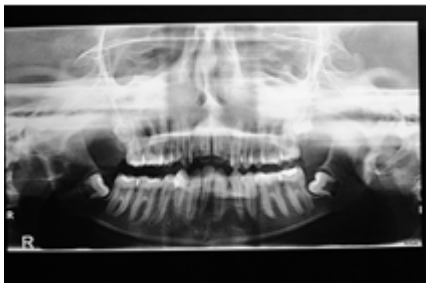

FIG 3(a) Orthopantogram

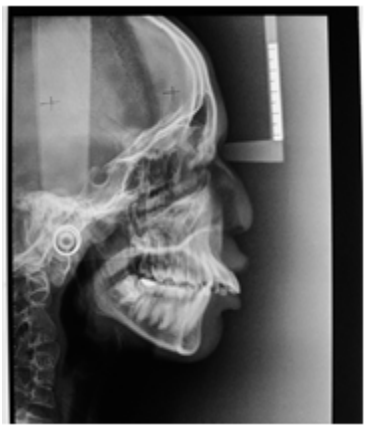

FIG 3(b) lateral cephalogram

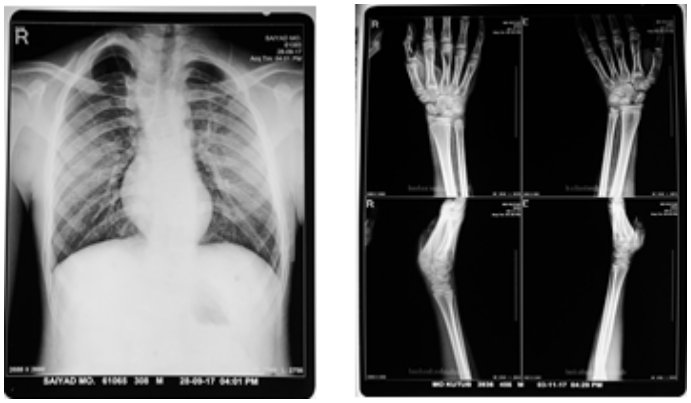

Fig 4 chest x- ray and x-ray of extremities.

DISCUSSION

Hallermann- Streiff syndrome is an uncommon congenital anomaly featuring oculo-mandibulo-cranial malformation. Diagnostic criteria by Francois[1] include 7 positive signs: 1)Dyscephalia and bird face 2)Proportioned dwarfism 3)Dental anomalies of number, position and shape 4) Hypotrichosis 5) Cutaneous atrophy, above all on the nose 6) Bilateral microopthalmia 7) Bilateral congenital cataract: total or partial and 5 negative signs that are certainly absent including, 1)No auricular anomalies 2)No palpebral anomalies 3)No muscular anomalies 4)No nails' and limbs' anomalies 5)Slight or no mental retardation[1]

The patient exhibited severe dentofacial characteristics that could be helpful in detailing the generic terms of "discephaly and bird face" as indicated by Francois. These signs can becharacterized as follows:

Craniofacial signs- zygomatic-maxillary hypoplasia, hypoplasia of the nose, which is small, sharpen, and beak-shaped, mandibular hypoplasia, with underdeveloped mandibular body and ramus and excessive width of the gonialangle.

Dental Anomalies- include natal and neonatal teeth, supernumerary teeth, malformed teeth, enamel hypoplasia, caries, malocclusion, agenesis of permanent teeth and delayed eruption of existing teeth.

Ocular Signs - microopthalmia with bilateral total or partial congenitalcataract.

Systemic Signs- proportioned dwarfism, cutaneous atrophy, slight deficiency in psychomotor development and hypogonadism and cryptorchidism.

Differential Diagnosis: The typical HSS characteristics that were found in the patient described here allow for differential diagnosis with respect to other syndromes of first brachial arch and ectodermic dysplasia, especially when associated with a reduced volume of craniofacial structures. In progeria there is a more generalized cutaneous atrophy, and ocular alterations are absent. In oculo-dento-digital dysplasia the face is similarto HSS, but anomalies of fingers (such as syndactyly and absence of the phalanxes) are present. Inmandibulofacial dysostosis the appearance of the face is analogous to HSS, but there are typical ear anomalies.In Seckel syndromeor bird-face dwarfism there are differences in auricular deformities aswell.

From the analysis of the literature and the observation of the present case it has to be emphasized that craniofacial and dental signs may play a major role within HSS polymorphism. A detailed appraisal of craniofacial and dentoalveolar characteristics appears to be of valuable assistance in the diagnosis of thesyndrome.

There is no cure reported for HSS. Symptomatic management with special attention to ophthalmologic, dental and upper respiratory problems to be carried out; Predisposition to severe dental caries and other dental problems makes counseling imperative. As for treatment of the dentoskeletal anomalies associated with HSS, the therapeutic protocol usedin the patient presented here was designed to reach the orthodontic goals during the developmental ages. Upper airway obstruction causes difficulties in airway management which may result from small nares and glossoptosis secondary to micrognthaia and tracheomalacia. Early deaths of the patients with HSS due to respiratory challenges necessitate prompt intervention by an ENT specialist.

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