

Hallermann Strieff Syndrome

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Abstract: Hallermann streiff is a rare syndrome. It was described by hallermann in 1948 and later by Streiff in 1950. A 5 yr. old female child had reported with typical bird like face shows frontal bossing, skin atrophy, hypotrichosis, of scalp, eyebrow and eye lash's, micro ophthalmia, congenital cataract, short stature suggestive of Hallermann-streiff syndrome. The case was presented because of its rare occurrence in the literature.

Keywords: Bird like face, cataract, Hallermann-streiff syndrome, microphthalmia

I. Introduction

Hallermann-streiff syndrome is a rare genetic disorder characterized by mandibulo facial abnormalities described by Hallermann in 1948 and later by streiff in 1950¹. There are a very low number of cases reported so far. Hallermann streiff syndrome often presents with sporadic mutation^{2, 3} clinical features are bird like face, bilateral micro ophthalmia, congenital cataract, skin atrophy, dental abnormalities and hypotrichosis. Patients usually die due to respiratory tract infection

II. Case Report

A 5 yr. old female child reported to our hospital with chief complain of redness and dimension of vision. Her birth history showed normal full term vaginal pregnancy was uneventful, but second degree of consanguinity was reported between the parents. Her parents and younger brother showed no significant findings. On the time admission her age was 5yrs. 2months with a height of 83.2cm (less than 3rd percentile) and weight 13kgs (less than 3rd percentile) showing growth delay >. She had frontal bossing, bilateral micro ophthalmia, bird like face, scanty eye lash eyebrow and hairs on the scalp, skin showed atrophy. Respiratory sounds were normal but stridor was heard while sleeping, no cardiac and per abdominal abnormalities.

2.1 Test Findings

Blood reports are as follows

Wbc-8,400/mm³, Hb-11.2gm/dl, Hematocrite-33.2%, MCV-80, Thrombocytic count -312/mm³, Proteins -6.7g/dl, albumin-4.2g/dl, BUN -11.0mg/dl, creatinine-0.4mg/dl. Her skull X-Ray revealed developmental failure in mandible and two fold increase in craniofacial ratio. In ophthalmological examination- scanty eye brows and eye lashes was noted, horizontal nystagmus, slit lamp examination confirmed micro cornea, A-Scan showed a short optic axis I.e. 17.2mm in right eye and 16.2mm in left eye exhibiting micro ophthalmia

2.2 Treatment

She was advised to come for a regular follow up to see the amount of increase in cataract. Cataract surgery would be done in future but Intraocular lens fitting the child's eye could not be found because of the micro ophthalmia and might also lead to post-operative complication like glaucoma. As of now she was prescribed glasses.

A

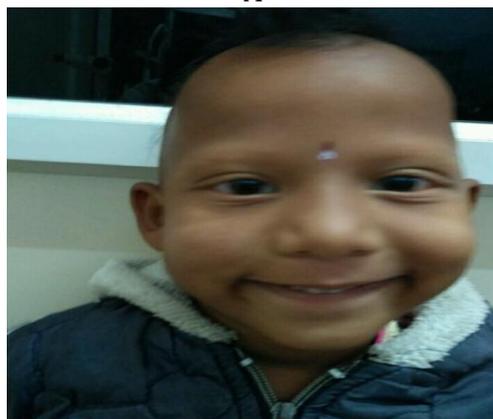




Fig 1a frontal bossing, bird like face, scanty eye brow and eye lash with micro ophthalmia 1b hypotrichosis of scalp

III. Discussion

Hallermann-Streiff syndrome, a rare congenital anomaly, was first reported incompletely by Aubry³) in 1893. In 1948, Hallermann⁴) made a complete report for the first time, which was followed by Streiff⁵) Report in 1950. In 1958 Francois⁶ analyzed clinical manifestation of 22 cases which has been described in various designation and sorted out 7 features 1-Dyscephalia and bird face 2- Dental anomalies ,3- proportionate nanism ,4- hypotrichosis ,5- Atrophy of skin ,6- Bilateral micro ophthalmia ,7- congenital cataract and reported under the name of “A New Syndrome”(Falls and schull⁷) named it ‘Hallermann-Streiff syndrome’ for first time as 6 cases were reported in 1960. In 1960 Steel and Brass¹⁰ analyzed 51 cases and included 1 case of their own . Nothing was known about the cause .It seemed that this syndrome did not have autosomal Dominant or Recessive type despite its sporadic manifestation. In beginning it was an assertion that it could be a mutation that occurred between 5th and 7th week of intrauterine life. Hypotrichosis is common and would be limited to cephalic region .body might show proportionate dwarfism .Developmental failure could not be due to hormonal deficiency .With respect to ocular findings bilateral micro ophthalmia and congenital cataract were manifested .Francois⁷ insisted that there was no mental disorder but Golomb and Poster⁹ showed mental disorder in 12 out of 80 cases .this cases did not show any mental retardation .A patient with Hallermann-Streiff syndrome would show complication due to anatomical structural abnormalities of upper respiratory tract like feeding problem, respiratory embarrassment, recurrent respiratory infection , sleep apnea. Endotracheal intubation could be difficult during general anesthesia .

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