EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 7
Issue: 10
546-548
Year: 2020

HALLERMANN STREIFF SYNDROME: A CASE REPORT

Dr. Ipsha Joshi*, Dr. Veena KH, Dr. Nisha*****

*Final Year PG Scholar Kle Shri Bmk Ayurveda Mahavidyalaya.
**Reader /Kle Shri Bmk Ayurveda Mahavidyalaya Karnataka.
***MD Kaumarabhritya Kle Shri Bmk Ayurveda Mahavidyalaya.

***Corresponding Author: Dr. Ipsha Joshi**
Final Year PG Scholar Kle Shri Bmk Ayurveda Mahavidyalaya.

Article Received on 20/08/2020 **Article Revised on 10/09/2020** **Article Accepted on 30/09/2020**

ABSTRACT
Hallermann Streiff Syndrome a rare disorder less then 200 cases reported having some peculiar features that are present since birth and will become more apparent over time. HSS (mandibulo occulofascial dyscephaly) is a branchial arch syndrome which combines with facial, dental and occular abnormalities, alopecia and may have unusual sutural distribution on scalp. A 4 year old boy presented with classical signs of Hallermann Streiff Syndrome. A thorough examination was performed. An interdisciplinary approach and preventive measures were taken regarding health of child.

KEYWORDS: Hallermann Streiff Syndrome.

INTRODUCTION

Hallermann Streiff Syndrome is a rare disorder and was first described in 1950. It is a rare disorder with less then 200 cases reported. The first description seems to have been made by Aubrey in 1893. HSS was first described completely in 1948 by Hallermann, and then in 1950 by Streiff.

HSS (mandibulo occulofascial dyscephaly) is a branchial arch syndrome which combines with facial, dental and occular abnormalities, alopecia and may have unusual sutural distribution on scalp.

CASE REPORT

A 4 year old boy brought by his uncle with chief complains of abnormally smaller eyes. Patient's history revealed that the parents had second degree consanguineous marriage. The baby was born on 9 months with prolonged normal vaginal delivery. He cried soon after birth but didn't opened his eyes for 6 hours and later had fever and was admitted in the hospital for 2 days for the same. Parents did not noticed any peculiar features regarding some abnormality at birth.

His medical history revealed delayed milestones, sleep apnoea, constipation .There was no history of mental retardation and was not under any medical intervention. The child was quite and cooperative during examination. According to guardian the child is fine in academics and is interactive with his age group and have friends.

On examination revealed facial characteristics like Bird face with pointed chin, micro-opthalmia and the distance between B/L inner canthus was 27.6mm, light brown coloured and sparse scalp hairs, hypotrichosis, mal implantation of teeth. The sparse hair on head, eyebrows and eyelashes were visible and were of similar light brown colour. The skull was brachycephalic with frontal bossing. A retrognathia and small mouth opening was present. Nutritional status of child revealed Ht for age was in less than 1 percentile, wt for age was between 1st to 3rd percentile and head circumference was between 3rd to 50th percentile. the visual acuity was 6/12.

.

Detailed oral hygiene instructions and dietary recommendations with a diet chart including per day calorie intake was provided at first appointment counselling to his guardian. The specialist opinion was taken regarding oral and dental hygiene and ophthalmological care. Cases of hallermann streiff syndrome are more prone to respiratory infections and these infections are also the most common cause of death in these cases. Early death of the patients with HSS due to respiratory challenges necessitates the improvement of quality of life.[2]

Detailed oral hygiene instruction was given to the child as well as the guardian. The regular visits will be required with an interdisciplinary approach and early preventive care programmes.

DISCUSSION

Seven essential signs were described by Francois as diagnostic criteria for HSS.[3]

Dyscephalia
Abnormal dentition
Bilateral micropthalmia
Hypotrichosis
Atrophy of skin especially over nose
Proportionate dwarfism
Congenital cataract

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. Physical growth and development was retarded however, mental development was normal in our patient. the dental hygiene was matter of concern including the malimplantation of teeth. The inheritance model is unknown and this syndrome seems to be a sporadic mutation. Familial cases have been reported. However, currently available data shows Inheritance of this developmental disorder is still unknown, and autosomal recessive as well as autosomal dominant inheritance with de novo mutations have been discussed.[4] Second degree Consanguineously married parents may have been one of the cause.[5] The consanguineous marriages being common in this part of the county could be one of the reason finding very rare genetic disorder that has only be identified only few times in the world. The education of people regarding the relationship between genetic disorders and consanguinity is the key if we want to stop such rare disorders to emerge in our population. In the rare front the quality of life must have been better for the patient if early diagnosis was made.

CONCLUSION

Support groups that can help to connect with other patients and families having the same condition, and provide valuable services are required. Developing the patient-centered information that can act as the driving force behind research for better treatments and possible cures. Educating the popuation regarding genetic disorders and consanguinity will play a important role in

preventing such rare disorders. Patients with Hallermann Streiff Syndrome face death due to respiratory challenges so regular checkups must be ensured. Proper dental hygiene and nutritional assessment is also very important. In the far end a multidisciplinary approach, parents and child councelling is necessary for betterment of patients suffering from Hallermann Streiff Syndrome.

REFERENCES

1. Thomas J, Ragavi B S, Raneesha P K, Ahmed N A, Cynthia S, Manoharan D, Manoharan R. Hallermann-Streiff syndrome. Indian J Dermatol, 2013; 58: 383-4.
2. Cho, Won-Kyung & Park, Joo & Park, Mi. Surgical Correction of Hallermann-Streiff Syndrome: A Case Report of Esotropia, Entropion, and Blepharoptosis. Korean journal of ophthalmology: KJO, 2011; 25: 142-5. 10.3341/kjo.2011.25.2.142.
3. Francois J. A new syndrome: Dyscephalia with bird face and dental anomalies, nanism, hypotrichosis, cutaneous atrophy, microphthalmia, and congenital cataract. AMA Arch Ophthalmol, 1958; 60: 842-62. [PUBMED]
4. Kortüm F, Chyrek M, Fuchs S, et al. Hallermann-Streiff Syndrome: No Evidence for a Link to Laminopathies. *Mol Syndromol*, 2011; 2(1): 27-34. doi:10.1159/000334317
5. pée E, Beleho D, Bitang AT, Njami VA, Bengondo C, Ebana Mvogo C. A familial study of Hallermann–Streiff–François syndrome. *Int Med Case Rep J*, 2017; 10: 193-201. https://doi.org/10.2147/IMCRJ.S114115.