Acta Scientific Ophthalmology

Volume 1 Issue 2 October 2018

Mini Case Study

Rare and Interesting Case of Goldners Syndrme in A 3 Years Old Male Child

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Received: September 20, 2018; **Published:** October 08, 2018

Abstract

It was in year 1850 an Austrian ophthalmologist by the name of Goldenhar was the first to describe a syndrome complex chartered by the cong presence of cong limbal dermoid with associated cog presence of preauricular skin tag R preauricular appendage. So, this entity became famous as go; Denhars syndrome sometimes presence of saint enophthalmos and CNG coloboma of u lid may also be seen. But it is very rare GHS is also termed as oculo auriculo vertebral syndrome or dysplasia with cranio facial dysplasia involving head and face in particular ear nose soft palate and mandible HSE associated cong anomalies in GHS occur only in 5 to 15 percent of cases and are due to incomplete development of 1st and 2nd branchial arch. Due to defect in genes not inherited autosomal dominant or recessive. Almost more than 80 percent case of GHS are normal as far as vision and mental faculty is concerned. Since 5 to 15 percent case have these additional cong lesions which are in the form of facial asymmetry high ached palate hare lip and cleft palate defects in kidney limbs and spine cong heart squint underlying orbital involvement defects in teeth and hearing impairment of mental faculty memory. So, we have to work up these cases and do following investigations 1: Ultrasound abdomen; 2: X-rays spine and limbs; 3: Echocardiography; 4: MRI orbits; 5: Complete eye examination; 6: Dental and ENT examination and 7: EEG.

Keywords: Cong Limbal; Dermoid Preauricular Appendage; Tag Facial Asymmetry; Hare Lip; Cleft Palate; High Arched Palate; Defects in Spine or Limbs; Cong Heart Defects in Teeth; Hearing Defect; Cong Heart Defects in Mental Faculty

Introduction

Coming to scenario of cong limbal dermoid they are usually unilateral may be bilateral. But very rare incidence is 1 in 10,000 or 1 in 3500 or 1 in 5,600 inferotemporal site is the commonest. About 70 percent male female ratio is 3 to 2 they may involve entire cornea or may be confined to conjunctiva only they are graded. According to corneal involvement grade 1, 2, 3; grade one is epithelial involvement, grade 2 is des membrane and grade 3 is involvement of entire ANT segment.

Case Report

3 years old male child was seen by me in my clinic with parents having noticed a small pale white inferotemporal limbal opacity of cornea. since birth left eye with associated presence of preauricular skin tag R side. This syndrome complex was characteristic of GHS child was born full term after LSCS. No history of exposure to oxygen or jaundice breath fed. Normal mile stones. No associated CNG anomalies vision mydriatic refraction, ANT SEG and fundii were normal. So, this child only needed counselling of parents. However, if the limbal dermoid involves pupillary are and threatens vision the modality of treatment is surgical which is both visual and cosmetic surgical. Procedures are 1: Lamellar keratoplasty, 2: Stem cell graft, 3: Amniotic membrane graft; 4: Smile lenticule tattooing and fibrin glue.

Discussion

GHS is a rare entity in most of case patients are normal as far as vision and mental faculty is concerned however only 5 to 15 percent case have additional cong anomalies [1-4].

Conclusion

Only these 5 to 15 percent case of GHS have too worked up for additional cong anomalies. otherwise we have to do proper counselling of parents if one gets a case of GHS without additional cong lesions.

Bibliography

1. We have GHS support groups.
2. There are families of GHS 17 such families of GHS are seen in Greece.
3. Children born in middle east during gulf war born in different military hospitals were reported to have high incidence of GHS.
4. Mittal., *et al*. "3 case of optic nerve drusen associated with GHS". *Indian Journal of Ophthalmology* (1968).

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