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Case Report

Rare Case of Goldenhar Syndrome in a 3 Years Old Male Child

Gowhar Ahmad*

Senior Consultant Ophthalmologist, Florence Hospital and University of Jammu and Kashmir, Jammu and Kashmir, India

***Corresponding Author:** Gowhar Ahmad, Senior Consultant Ophthalmologist, Florence Hospital and University of Jammu and Kashmir, Jammu and Kashmir, India.

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Abstract

Cong limbal dermoid preauricular skin tag appendage squint enophthalmos colobomas of upper lid iris retina astigmatism microphthalmos blepharophimosis syndrome facial asymmetry high arched palate hare lip cleft palate double ureters hydronephrosis defects in limbs and spine cong heart dental anomalies hearing defects impairment of memory intelligence mental facility 7th nerve involvement trigeminal anasthesia.

Keywords: Goldenhar Syndrome; Cong Limbal Dermoid; Squint Enophthalmos

Introduction

It was Maurice Goldenhar an Austrian ophthalmologist who in the year 1850 was the first to have described a syndrome complex characterized by the presence of cong limbal dermoid with associated cong presence of preauricular skin tag or preauricular appendage sometimes presence of squint enophthalmos coloboma of the UNLID iris retina astigmatism microphthalmos and blepharophimosis syndrome may also be seen however it is very rare.

GHS is also termed as oculo auriculo vertebral syndrome and cranio facial syndrome involving head face ear nose soft palate and mandible.

80 to 85 percent cases of ghs are normal from visual and mental facility point if view only 10 to 15 percent case have additional associated cong anomalies which are due to incomplete development of first and second branchial arch due to the defects in genes not hereditary autosomal dominant and recessive or may be due to maternal gestational d m or exposure to rubella haemophilus or cytomegalic Inc virus in the first trimester of pregnancy or history not intake of thalidomide cocaine and retinoic acid.

These additional cong anomalies are in the form of facial asymmetry high arched palate hare lip cleft palate.

Defects in limbs and spine kidney lesions like double ureters hydronephrosis cong heart underlying orbital involvement dental anomalies hearing defect impairment of mental facility memory.

7th nerve involvement aplasia of trigeminal nuclei trigeminal anasthesia.

Someone has to work up these anomalies and following investigations

- Ultrasound abdomen for kidney defects
- X-rays spine and limbs
- MRI orbits to exclude underlying orbital involvement
- Echocardiography for cong heart
- Dental examination
- Audiometry for hearing defects
- EEG.

As far as scenario of limbal dermoid is concerned.

They are usually unilateral can be bilateral but very rare may involve entire cornea or may be only confined to conjunctiva.

Incidence is v1 in 10.000.

Inferotemporal site is the commonest about 70 percent.

Male female ratio is 3 to 2.

They are graded according to corneal involvement:

- Grade 1 is involvement of corneal epithelium
- Grade 2 is involvement of Descemet's membrane
- Grade 3 is involvement of entire ant segment.

Case Report

3-years old male child was seen by me some time ago in my office with non vision married couple having noticed a small palish white lesion at the inferotemporal site r eye since birth with associated presence of preauricular skin tag this was characteristic of ghs.

Ft delivered male child after LSCS.no history of exposure to oxygen or jaundice normal mile stones breast fed vision ant segment mydriatic refraction and fundii normal no associated cong anomalies.

So, this child had limbal dermoid at the commonest site not involving visual axis to threaten vision needed only reassurance forte parents and observation.

However, if we have limbal dermoid involving visual axis to threaten vision treatment is surgical which is both

- Visual and
- Cosmetic
- Surgical modalities
- Lamellar keratoplasty
- Autograft
- Stem cell graft
- Amniotic membrane graft
- Smile lenticule tattooing fibrin glue.

Here we exercise limbal dermoid subject to tattooing to maintain colour and later corneal lenticule is put in situ with fibrin glue as no stiches are applied so post-operative pain and astigmatism is less.

Discussion and Conclusion

Ghs is a Cong genetic condition characterized by cong presence of limbal dermoid and cong preauricular skin tag or preauricular appendage.

However, 80 to 85 Percent cases are normal from visual and mental facility point of view it is only in 10 to 15 percent of cases we have additional cong anamolies for which special care has to be taken.

Reassurance to parents and observation is necessary [1-5].

Conflict of Interest

I have no financial interest in publishing this article.

Bibliography

1. We have GHS support groups.
2. Their are families of GHS.
3. 17 such families are seen in Greece.
4. “Children born in middle East during gulf war born in different military hospitals had high incidence of GHS”.
5. Mittal., *et al*. in the year 1968 reported 3 cases of optic nerve drusens associated with G H S.