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Short Communication

Name Rare and Interesting Case of Goldenhar's Syndrome in a 3 Years Old Male Child

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Credit of GHS goes to an Austrian ophthalmologist Maurice Goldenhar who in the year 1850 was the first to describe a syndrome complex characterized by cong presence of limbal dermoid with cong associated presence of a preauricular skin tag or appendage sometimes presence of squint anophthalmos coloboma of u lid iris.

Retina astigmatic ref error microphthalmos or blepharophimosis.

Syndrome may be also seen however it is very rare. GHS is also termed as occulo auricula vertebral syndrome or dysplasia and craniofacial Syndrome or dysplasia involving head and face ear nose soft palate and mandible. These dysplastic lesions are responsible for additional anomalies in GHS however they occur only in 5 to 15 percent case ad are due to incomplete development of 1st and 2nd branchial arch due to defect in genes not inherited autosomal dominant and recessive may be due to maternal gestational DM or exposure to rubella and influlanza virus or history of drug in to cocaine thalidomide retinoic acid antibiotics.

The additional cong anomalies may be facial asymmetry high arched palate hare lip cleft palate defects in kidney hydronephrosis double ureter defects in limbs spine cong heart underlying orbital involvement dental anomalies hearing defect impairment of memory CNS involvement 7th unilateral aplasia of trigeminal nerve and trigeminal anasthesia.

However 80 to 85 percent cases of GHS are normal from visual and mental facial point of view.

Dermoid are usually unilateral can be bill but rare inferotemporal site is the commonest about 70 percent incidence is 1 in 10 000 may involve entire cornea or may be confined to conjunctiva only male female ratioi is 3 to 2.

Graded according to corneal involvement

- Grade 1 epithelial involvement
- Grade 2 descemet's membrane involvement
- Grade 3 involvement of whole of at segment.

Case Report

Few years back a 3 years male child was seen by me with parents having noticed inferotemporal limbal site a plash lesion since birth r eye with cong presence of a preauricular skin tag R ear this was a case of GHS child had no other associated cong defects VA ref ant and post seg refraction and fundii were normal FT child born after l s c s breast fed normal mile stones.

This child only need reassurance for the parents. however if we have a case where limbal dermoid involves visual axis and threatens vision one has to do surgery which is both cosmetic visual.

They are

- Lamellar keratoplasty
- Autograft
- Stem cell graft
- Ammonitic membrane graft
- Smile lenticular tattooing and fibrin glue
- References

We have GHS support groups. Their are 17 families of GHS in Greece. Children born in middle east during gulf war born in different military hospitals had high incidence of GHS. Mittal., *et al*. in IJO 1968 reported 3 cases of drusen's associated with GHS.