Abstract

RARE CASE OF GOLDENHARS SYNDROME IN A 3 TEARS OLD MALE CHILD

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INTRODUCTION

It was Maurice goldenhar an Austrian ophthalmologist who in the year 1850 was the first to describe a Syndrome complex chareterised by the presence of cong LImbal dermiod with assiciated cong presence of Preauricular skin tag or Preauricular appendage sometimes presence of squint Anophthalmos Coloboma of the u lid iris retina astigmatism MICROPHTHALMOS and blephrophymosis synrome may be seen however it is very rareG h s is also termed asOcclulo auriculo vertebral Syndrome
And craniifacial Syndrome involving head face ear nose soft plate and mandible80 to 85 percent cases of g h s are normal from.visual and Ophthalmic point of view.It is only in 10 to 15 Percent cases that we have got additional cong defects which are due to incomplete Devolpment of ist and 2nd branchial arch due to defects in genes not inherered autosomal dominant and recessive or could be due to maternal gestational d m or history of intake of thalidomide cocaine and retinoic acid or exposure to rubella heamophilus
The additional cong defects are facial asymetry high arched palate hare lip cleft palate defects in kidney defects in limbs and spine
Cong heart hearing defects dental Anamolies impaiment of memory. So we should work up the cases and do xrays spine limbed ultrssound abdomen echocardiogram dental and e n t EXAMINATION.
MRI orbits eegAs far as scenrio of cong Dermiod is concerned they are unilateral can be bil very rare they may involve entire cornea or may be only confined to Conjuctiv.Prevelance is 1 in 10 000Inferotemporal site is the commonest 70 percent Graded according to the corneal involement
Grade 1,Grade 2, Grade 3
Grade 1 is corneal epithelial involement
Grade 2 is involement of the rest membrane.
Grade 3 is involement of entire ant segment.

Case report
I happen to see a 3 years old male child in my office some time ago with parents having noticed a palish white lesion on the inferotemporal site of Limbus r eye since birth with assiciated cong presence of Preauricular skin tag on left side this syndrome complex was characteristic of g h s
The child was born full term from non cousin married parents following LSCS no history of exposure to oxygen or jaundice normal mile stones and brest fedVision refration and fundii normal no other assiciated cong defects.Grade I cong LImbal dermiod not involvimg visual axis treatment is Parenteral counseling and observation. However if LImbal Dermiod involves Visual axis to thereten vision treament is surgical which is both Visual and Cosmotic, Modalities of surgery, Lamellar kerataplasty.

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