
Abstract

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

Dr gowhar Ahmad

INTRODUCTION

It was Maurice Goldenhar an Austrian ophthalmologist who in the year 1850 was the first to describe a Syndrome complex characterized by the presence of congenital limbic dermoid with associated congenital presence of Preauricular skin tag or Preauricular appendage sometimes presence of squint Anophthalmos Coloboma of the upper lid iris retina astigmatism MICROPTHALMOS and blepharophymosis syndrome may be seen however it is very rare Goldenhar's is also termed as Oculo auriculo vertebral Syndrome

And craniofacial Syndrome involving head face ear nose soft palate and mandible 80 to 85 percent cases of Goldenhar's are normal from visual and Ophthalmic point of view. It is only in 10 to 15 Percent cases that we have got additional congenital defects which are due to incomplete development of first and second branchial arch due to defects in genes not inherited autosomal dominant and recessive or could be due to maternal gestational damage or history of intake of thalidomide cocaine and retinoic acid or exposure to rubella haemophilus

The additional congenital defects are facial asymmetry high arched palate hare lip cleft palate defects in kidney defects in limbs and spine

Congenital heart hearing defects dental Anomalies impairment of memory. So we should work up the cases and do xrays spine limb ultrasound abdomen echocardiogram dental and ENT EXAMINATION.

MRI orbits EEG As far as scenario of congenital Dermoid is concerned they are unilateral can be bilateral very rare they may involve entire cornea or may be only confined to Conjunctiva. Prevalence is 1 in 10 000 Inferotemporal site is the commonest 70 percent Graded according to the corneal involvement

Grade 1, Grade 2, Grade 3

Grade 1 is corneal epithelial involvement

Grade 2 is involvement of the rest membrane.

Grade 3 is involvement of entire anterior 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 of eye since birth with associated congenital presence of Preauricular skin tag on left side this syndrome complex was characteristic of Goldenhar's

The child was born full term from non-cousin married parents following LSCS no history of exposure to oxygen or jaundice normal milestones and breast fed Vision refraction and fundus normal no other associated congenital defects. Grade I congenital Limbal dermoid not involving visual axis treatment is Parenteral counseling and observation. However if Limbal Dermoid involves Visual axis to threaten vision treatment is surgical which is both Visual and Cosmetic, Modalities of surgery, Lamellar keratoplasty.

Dept of ophthalmology Florence hospital Chanapora Srinagar Kashmir, India

Correspondence: Dr gowhar Ahmad, Dept of ophthalmology Florence hospital Chanapora Srinagar Kashmir, India E-mail:gowhar.ahmad1948@gmail.com



This open-access article is distributed under the terms of the Creative Commons Attribution Non-Commercial License (CC BY-NC) (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits reuse, distribution and reproduction of the article, provided that the original work is properly cited and the reuse is restricted to noncommercial purposes. For commercial reuse, contact reprints@pulsus.com

