

An Interesting Case of Golden Hars Syndrome in 3 Years Male Child



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Abstract

Golden hars syndrome is a syndrome complex characterized by congenital presence of a limbal dermoid with congenitally associated presence of pre auricular skin tag or pre auricular appendage sometimes there may be presence of squint the condition is called as ocular auricular vertebral syndrome or dysplasia the defect is in genes it is not inherited due to incomplete development of 1st and 2nd bronchial arch some genetic defects in head and face called as cranio facial dysplasia involving head and face common organs involved are 1 ear 2 nose 3 soft palate and mandible it occurs only in 5 to 15 percent of cases so one gets defects in 1 spine 2 limbs 3 kidney 4 hearing problem 5 dental anomalies 6 congenital impairment of memory so one should do following investigations in these cases

1. x-ray limbs and spine
2. Ultrasound abdomen
3. Echo cardiography
4. Dental examinations
5. MRI orbits to exclude underlying orbital involvement.

Keywords: Ocular auricular vertebral syndrome; Cranio facial dysplasia; Limbal dermoid; Squint; Congenital heart; Dental anomalies; Defects in limbs and spine; Squint

Introduction

Now we have defined Golden Hars Syndrome as a complex of congenitally associated limbal dermoid with congenitally associated pre auricular skin tag or appendage. Limbal dermoids sometimes are bilateral rarely. Usually they are unilateral involving whole of cornea or may be confined to conjunctiva only. Incidence of limbal dermoids are 1 in 10000 or 500 in 2700 they are graded according to involvement of cornea grade 1. when only corneal epithelium is involved 2 grade 2 Descemet membrane 3 grade 3 whole of anterior segment of eye is involved. Inferotemporal involvement of limbal dermoid is the commonest about 70 percent.

Case Report

6 months back a 3 years male child was seen by me in my office with parents having noticed a small whitish. White inferotemporal limbal region left eye with congenitally associated presence of pre auricular skin appendage. Child delivered after LSCS no other

congenital anomalies seen. Vision mydriatic refraction anterior segment and fundi were normal. Child had grade 1 limbal dermoid with the commonest site being inferotemporal.

Results and Discussion

Golden Hars is not a vision threatening disease most of these patients have normal intelligence and vision only in cases where visual axis is involved. We think of surgery. Conclusion: My case was a simple case of Gowhar Ahmad grade 1 limbal dermoid so it only needs observation in cases where visual axis is involved and vision is threatened then the surgical modalities are

1. Visual
2. Cosmotic

We do

1. Lamellar keratoplasty

2. Amionotic membrane graft
3. Stem cell graft

Their families of golden hars syndrome in Greece 2 in middle east in gulf war children born in different military hospital had golden hars syndrome 3 sometimes optic nerve drusen is associated with g h syndrome.

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