

Goldenhar syndrome: Dermoid excision reduces acquired astigmatism

Amit Mohan, M.S.; Sudhir Singh, M.S.

Correspondence Author
Dr Amit Mohan, M.S
Global Hospital Institute of Ophthalmology,
Abu Road, Sirohi
Rajasthan 307501

Abstract

Goldenhar syndrome is a rare congenital anomaly which consist of a triad of an ocular dermoid cyst, preaural skin tags and vertebral dysplasia. We report a case of Goldenhar syndrome diagnosed in a 19yr old girl with acquired astigmatism of 3 diopters against the rule and decreased to 1 diopter against the rule asigmatism after dermoid excision.

Case Report

A 19 year old girl presented with congenital lesions of the face and eyes. Ocular examination revealed a dermoid nodule located on the bulbar conjunctiva at inferotemporal side of limbus in the Left Eye (fig-1).



Fig.1 A dermoid nodule located on the bulbar conjunctiva at inferotemporal side of limbus in the Left Eye

Refraction identified against the rule astigmatism of 3 diopters. BCVA was 6/6 in right eye and 6/18 in left eye. Ocular motility was normal.

On facial examination there was a preaural tag near the tragus of pinna (fig.-2)



Fig.2. Showing pre aural tag near tragus of the pinna of the left eye.

Skeletal examination showed micrognathia and dental anomalies (fig-3)

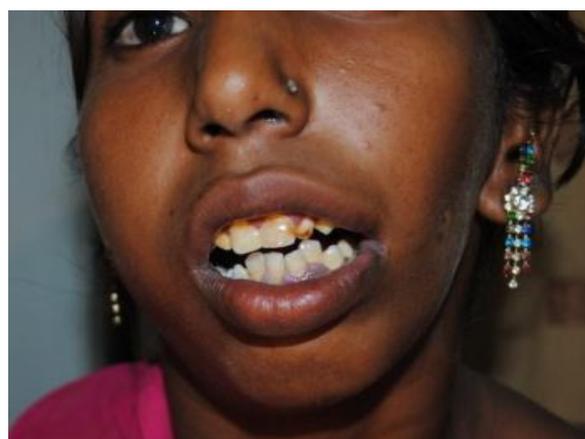


Fig.3 Showing micrognathia and dental anomalies

which was confirmed in x-ray film (fig-4 & 5). These symptoms were consistent with the diagnosis of Goldenhar syndrome.



Fig 4 X ray film showing dental abnormality



Fig 5. Fig 4 X ray film showing micrognathia

Complete excision of dermoid cyst was done(fig-6).



Fig.6 Appearance after dermoid excision

Intraoperative and postoperative course was uneventful. Postoperative Astigmatism reduced to 1 diopter against the rule astigmatism.

Discussion

Goldenhar Syndrome also known as oculoauriculovertebral syndrome is a rare congenital malformation involving the first and second branchial arches ¹. It was first described by Goldenhar in 1952. If the limbal dermoid are detected and excised early astigmatism may be reduced. The main ocular feature of Goldenhar syndrome is epibulbar choriostoma in 30 to 60 %^{2,3}. It consist of a dermoid or lipodermoid. The corneal and the sclera invasion by the tumour is rare and leads to against the rule astigmatism. Management of ocular dermoid is surgical excision. Dermoid cyst is a benign tumour that causes serious ophthalmological sequels-Astigmatism, Amblyopia &Strabismus. The ophthalmologist should focus on visual consequences, treat early and follow up the patient because dermoid causes acquired astigmatism and amblyopia.

References

- 1) Bijal M, Nayak S, Shankar S- Goldenhar syndrome with unusual features, *Indian –J.Dermatol*,2008;74 254-256.
- 2) Anderson PJ, David DJ-Craniofacial J.2005; 42 477-80.
- 3) Murat O, Mesut G, Aylekin G, Guven O –Goldenhar syndrome associated with bilateral ocular choriostoma and cardiac abnormalities, *Eur J Gen Med* 2004; 1: 28-30.