



## A Case Report of Goldenhar Syndrome With Unilateral Peter's Anomaly

### KEYWORDS

Goldenhar's Syndrome, PETER's Anomaly, Lid Coloboma, Corneal opacity, Limbal Dermoid

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**ABSTRACT** We are reporting a Case of Goldenhar Syndrome with unilateral PETER's anomaly which is a rare entity. Our case details are a 12 years boy presented with Defective vision in both eyes for distance and near. Ocular lesions : right eye - Small gap in the upper eye lid lower margin. A small white opacity in the right para central area of the cornea. A growth in the left eye near limbus measuring 3x3mm circular, Ear deformitis - small multiple skin tags in front of tragus on both sides of ears, Skin lesion - A small hypo pigmented patch measuring 7x8mm on the right hand fore arm. Mild right side deviation of the vertebral column seen. Lid coloboma and pre Auricular skin tags were rectified with the help of plastic surgeon. A single case of this type with little variation has been reported from AIIMS, India. And this is the second case with above unique features. This case may be first of its kind both from India as well as from Western World.

### Case Report :

A male 12 years boy presented with Defective vision in both eyes for distance and near. Ocular lesions : right eye - Small gap in the upper eye lid lower margin. A small white opacity in the right para central area of the cornea. A growth in the left eye near limbus measuring 3x3mm circular, Ear deformitis - small multiple skin tags in front of tragus on both sides of ears, Skin lesion - A small hypo pigmented patch measuring 7x8mm on the right hand fore arm. Mild right side deviation of the vertebral column seen. Lid coloboma and pre Auricular skin tags were rectified with the help of plastic surgeon.

**Family History:** Consanguineous marriage

**On Examination :** Skin lesion: Tags in front of Ear, bilateral, Hypo pigmental patch on fore arm. Ocular examinations - Vision: 6/12 6/6, No refractive errors, **Anterior segment** - Right Eye-Coloboma of upper lid 8x4-near medial conthus, Corneal opacity (leuco matus)-3x3mm paracentral, Irido corneal adhesions (strands) at center of cornea to the anterior surface of iris, Left Eye-Dermoid measuring 3x3mm at limbus 4 to 5 o'clock. Both eyes funds and intraocular pressure were found to be normal. All Other systems found to be Normal. With the above findings we have made a diagnosis of Goldenhar Syndrome with unilateral PETER's anomaly.

**Discussion:** Combination of Goldenhar Syndrome with unilateral PETER's anomaly is a rare entity not reported very frequently. Our case is of unique type which was not reported as such in the literature. Patients right eye lid coloboma and pre auricular skin tags were rectified surgically with the help of plastic surgeon. Corneal opacity in right eye is small and para central not effecting the vision much and hence left untreated. Limbal Dermoid in left eye not causing any symptoms and hence not treated. Cause for drop in right eye visual acuity (6/12) has been attributed to corneal opacity in the papillary area.

Goldenhar syndrome also known as Oculo-Auriculo-Vertebral (OAV) syndrome/ hemifacial microsomia is a rare con-

genital defect characterized by incomplete development of the ear, nose, soft palate, lip, and mandible. It is associated with anomalous development of the first branchial arch and second branchial arch. Common clinical manifestations include limbal dermoids, preauricular skin tags, and strabismus. Maurice Goldenhar has reported this case in 1952. The cause of Goldenhar syndrome is not known but there may be a genetic component, with multy factorial inheritance. It is also suggested that there is a defect in branchial arch development in the first trimester.

Peters anomaly is a rare form of anterior segment dysgenesis in which abnormal cleavage of the anterior chamber occurs. Involving the central or entire cornea. In our case Peters anomaly is of type one. Peters anomaly is may also be associated with myopia, aniridia, coloboma of the iris, choroid, microphthalmos, persistent hyperplasia of primary vitreous (PHPV), and optic disk hypoplasia.

**Conclusion:** we are reporting a successfully managed case of combination of Goldenhar Syndrome with unilateral PETER's anomaly type -1.

### Pictures of Patient before and After surgical treatment:



Before Surgical Correction



After Surgical Correction