ORIGINAL RESEARCH PAPER
Volume-6 | Issue-11 | November-2017 | ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

LIMBAL DERMOID EXCISION WITH AMNIOTIC MEMBRANE GRAFTING IN GOLDENHAR SYNDROME

Ophthalmology

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KEYWORDS

INTRODUCTION:
Goldenhar syndrome is also known as oculo-auriculo-vertebral dysplasia with hemifacial microsomia which is a rare congenital anomaly involving the first and second branchial arches.The syndrome was first described in 1952 by the French ophthalmologist Maurice Goldenhar.The incidence of Goldenhar syndrome has been reported to be between 1:3500 and 1:5600, with a male:female ratio of 3:2. The exact etiology is not known .Most of the cases have been noted as sporadic. Ingestion of drugs such as thalidomide, retinoic acid, tamoxifen, and cocaine by the pregnant mother may be related to the development of this syndrome.

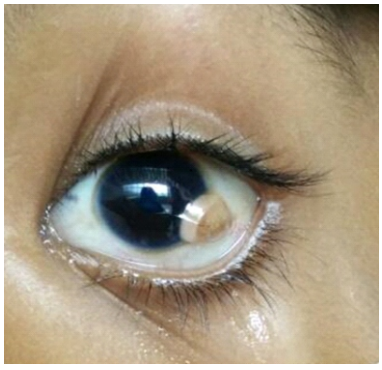
CLINICAL CASE REPORT:
12 year old male patient, presented to tertiary eye hospital , on 11/11/15 with complain of ocular discomfort and swelling in the left eye for 1 year.The swelling was gradually increasing in size, for which the patient has consulted a private practitioner and was advised lubricating and antibiotic eyedrops .There was past history of pre-auricular tags which was surgically removed in a local hospital 3 years back. (there was no other significant records available.)

Birth history:The child was born of non consanguineous parents at full term normal vaginal delivery and without any perinatal complication.Mother of this patient was not taking any kind of drugs during pregnancy.No significant family history was present.

ON EXAMINATION:
Vitals were within normal limits.
On general examination , scoliosis of spine and aural tags were noted.

ON OPHTHALMOLOGICAL EXAMINATION:
Both eye best corrected visual acuity was 6/18. Intra Ocular Pressure measured by Perkins Tonometer was 12 & 14 mm Hg respectively. On Anterior segment examination in left eye revealed a 10 *8 mm in size, oval , well circumscribed limbal swelling in the inferotemporal quadrant, enchroching 3 mm on the corneal surface,Which was yellowish in colour and having smooth surface and well defined border. Posterior segment evaluation revealed healthy disc, healthy orange fundus glow, normal Cup :Disc ratio and dichotomously branching vessels in both eyes.

Right Eye		Left Eye
Normal	Lids	Normal
Normal	Lacrimal Apparatus	Normal
Normal	Conjunctiva	Limbal Dermoid Present
Clear	Cornea	Clear
Normal Depth	Anterior Chamber	Normal Depth
Normal Reacting To Light	Iris /pupil	Normal
Clear	Lens	Clear
Orange	Glow	Orange



METHOD:
From clinical presentation and examination , limbal swelling was diagnosed as limbal dermoid .So, surgical excision of the limbal dermoid followed by amniotic membrane graft with fibrin glue was planned.Routine systemic examination and pre operative work up was performed.All blood investigation were within normal limits.

The surgery was performed under general anesthesia :
Surgical excision of the dermoid was performed.Thorough hemostasis achieved.The underneath bare sclera was measured.Appropriate sized amniotic membrane excised and placed over bare sclera and secured with help of fibrin glue.No sutures were taken.and eye was carefully patched taking care not to displace the graft. And speciment was sent for histopathological examination.

Post operative treatment:
Lubricating eyedrops 6 times/day, Antibiotic eyedrops (Gatifloxcacin & Tobramycin) 6 times/day, Low dose steroids (Flurometholone) 4 times/day tapered off every 5 days, were prescribed.

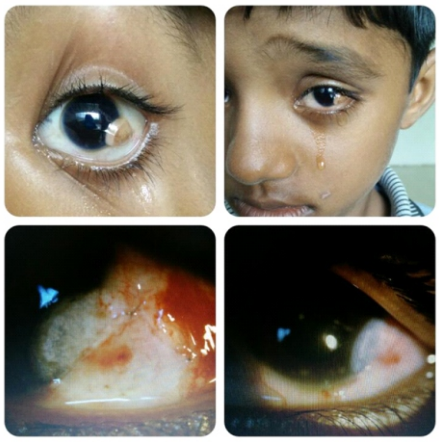
RESULT:
Post op follow up was taken at day 1, day 5, 2 week, 1, 3 months.

On day 1: The amniotic membrane graft was in place. There was ciliary congestion and an area of sub conjunctival haemorrhage at 7 o'clock, which cleared up by 2 weeks.

At the end of 1 months, the amniotic membrane graft had completely settled and acquired transparency, with no evident vascularisation or inflammation.

At the end of 3 months, there was no more ocular discomfort or any recurrence of a similar swelling.

Histo-pathological reports confirmed the lesion to be a solid limbal dermoid. The tissue revealed stratified squamous epithelium with thin keratinisation superficially, with stroma containing disordered pilar structures, inflammatory exudates, vascular bodies and some amount of adipose tissue, ruling out any malignant changes.



DISCUSSION:

The classic features of this syndrome include ocular menifestations such as microphthalmia, epibulbar dermoids, lipodermoids, coloboma; Aural features such as pre-auricular tragi, hearing loss, Microtia.Vertebral anomalies such as scoliosis, hemivertebrae, and cervical fusion.Limbal dermoids are usually recurrent and are conventionally treated with dermoid excision with bare sclera technique or conjuctival autograft.But in our case we have attempted surgical excision of limbal dermoid followed by amniotic membrane grafting with fibrin glue.

The amniotic membrane is a derivative of the fetal ectoderm that makes up the innermost layer of the fetal membrane. It was first used as a substitute for rabbit peritoneum [1] in the management of ocular chemical burns and now has a varied amount of uses, including its use in recurrent corneal erosion and severe dry eye. Amniotic membranes are being used as grafts due to their ability to reduce postoperative pain, corneal neovascularization and inflammation that leads to scarring due to its in-vitro inhibition of TGF B [3] . They also promote epithelialization and differentiation of ocular epithelium by protection of limbal stem cells [1] . For these reasons, we used the amniotic membrane as our graft of choice. Extra caution was taken in our case by using a fibrinogen plus thrombin glue which forms a smooth seal along the wound edge and dissolvable 9.0-vicryl sutures to ensure proper placement and fixation of the amniotic graft in this pediatric patient.

Mahdavi and Pourafkari reported a case of a 19-year-old man with a limbal dermoid cyst measuring 5 mm by 6 mm with black hair. The lesion was excised, and lamellar keratoplasty performed. They saw minimal improvement in visual acuity after surgery as a result of the amblyopia and induced astigmatism [5] . Pirouzian et al. reported a case of three limbal dermoid patients who underwent deep lamellar excision followed by sutureless multilayered amniotic membrane transplantation and reported improved cosmetic outcomes [4] . Lazzaro describes the excision of a limbal dermoid with the placement of a processed pericardial graft and stated beneficial results for larger lesions [5] .

Complications after surgery may include potential risk of scarring, loss of vision, and development of pseudopterygium. However, use of amniotic membrane may reduce the regrowth of the dermoid needing repeat surgeries in our experience. Lang et al. purported that the transplantation of amniotic membrane following removal of a limbal dermoid could not prevent the occurrence of a pseudopterygium but the use of Mitomycin C had a protective effect. Although vision improved slightly, we did not notice a significant change in astigmatism or anisometropia with our patient, consistent with other studies [6] .

Using the amniotic membrane after excision of the dermoid is a relatively new technique. A combination of the amniotic membrane and use of the thrombin/fibrinogen glue gave us a slight improvement in vision not seen with some techniques but the degree of astigmatism remained unchanged. More work needs to be done to develop a technique that improves the degree of astigmatism after excision of limbal dermoids but we propose that surgery should be considered over medical management in a case of persistant discomfort, disfigurement, visual changes and an enlarging cyst.

Conclusion:

This case has been presented to increase the awareness about this rare entity, to highlight the typical clinical and histo-pathological findings & treatment approach using amniotic membrane graft modality in cases of dermoid associated with Goldenhar syndrome.

Limbal dermoids are usually recurrent and are conventionally treated with dermoid excision with bare sclera technique or conjuctival autograft.

The long term advantages of Amniotic membrane grafting include graft stability, better cosmesis and relatively less chances of recurrence.

And like this case if the graft is used with a fibrin glue, suture placement can be avoided, thereby decreasing the postoperative foreign body sensation and discomfort & increasing patient compliance and outcome

References:

1. Kanski clinical ophthalmology 8th edition page no. 669-672.
2. Riyaz A, Riyaz N. Goldenhar syndrome with unusual features. Indian J Dermatol Venereol Leprol.
3. Kulkarni V, Shah MD, Parikh A. Goldenhar syndrome: A case report. J Postgrad Med.
4. Reddy MV, Reddy PP, Usha Rani P, Hema Bindu L. Facio-auricular vertebral syndrome: A case report.Indian J Hum Genet.
5. Kapur R, Kapur R, Sheikh S, Jindal S, Kulkarni S. Hemifacial microsomia: A case report. J Indian Soc Pedod Prev Dent.
6. Miller TD, Metry D. Multiple accessory tragi as a clue to the diagnosis of the oculo-auriculo-vertebral (Goldenhar) syndrome. J Am Acad Dermatol.