

Presentation and Surgical Management of Epibulbar (Limbal) Dermoids

Khawaja Khalid Shoaib, Tariq Shakoor, Muhammad Shahbaz Amin

Pak J Ophthalmol 2018, Vol. 34, No. 3

See end of article for authors affiliations

Correspondence to:
Khawaja Khalid Shoaib
Health Bridge Hospital
Ghazi Road, Near Bhatta
Chowk, DHA, Lahore
Email: kkschoaib@hotmail.com

Purpose: To report clinical characteristics and surgical outcomes of limbal dermoids excision in Pakistani patients.

Study Design: Retrospective, Descriptive.

Place and Duration of Study: Mughal Eye Hospital Lahore from 1st June 2016 to 30 Dec 2017.

Material and Methods: Records of 15 epibulbar dermoids were reviewed. Site, colour, presence of pigmentation and presence of ocular and systemic associations of the lesions were studied. Lesions were photographed and then surgically managed. Excision was done with blade and scissors. Postoperative follow up included management of complications and photograph on each visit.

Results: Age ranged from 4 to 60 years (Mean 18 ± 13.48). Male to female ratio was 6:9. All the patients had unilateral epibulbar dermoids, which were present in inferotemporal quadrant of the limbus. Most lesions (9 cases - 60%) involved cornea and sclera equally while a few extended more on the scleral (3 cases - 20%) or corneal side (3 cases - 20%). Most (12 cases - 80%) were round and a few (3 cases - 20%) tapering. 5 (33%) had Goldenhar Syndrome. 4 (27%) patients had preauricular tags. One (7%) patient had maxillary hypoplasia and divergent squint. Postoperatively one patient (7%) had corneal thinning and one patient (7%) had extensive formation of granulation tissue. There was pigmentation of the lesion in 12 cases (80%). Three (20%) patients had microtia.

Conclusion: Limbal dermoids in Pakistani patients have characteristics resembling those described in other parts of the world. Treatment with excision and superficial sclerokeratectomy without graft gives satisfactory results.

Keywords: Epibulbar Dermoids, Limbal Dermoids, Goldenhar syndrome.

E pibulbar dermoid is a choristoma. It is composed of fibrous and fatty tissue, covered by keratinized epithelium. It is present from birth. A few produce a lipoid infiltration of the corneal or scleral stroma at their leading edge. In some instances there may also be subconjunctival dermolipomas (adipose tissue and dense connective tissue) which are present in the lateral quadrant of the eye. These can be up to 10 mm in diameter and usually straddle the limbus. Most are on the inferior temporal limbus^{1,2}. The pathogenesis of dermoids is multifactorial. Very rarely more than one family

members have been found to have similar lesions³. On histopathological examination, they contain many tissues including skin, hair, fat, sweat gland, connective tissue, lacrimal gland, muscle, bone, teeth, cartilage, vascular/neurologic tissue and may even contain brain tissue. Lymphoid elements can also be present.

Dermoids are classified into three types on the basis of location of the lesion. The most common involves the limbus. Limbal dermoids mostly present as superficial lesions but deeper ocular structures can also be involved. The second type is entirely in the

superficial cornea. The last variety of dermoid is rare and affects full thickness of cornea and deeper tissue are replaced with a fibrous and fatty tissue.

Epibulbar Dermoid is often seen with Goldenhar (oculoauriculovertebral) syndrome⁴. These patients may have a variety of other anomalies, including ear deformities (partially formed ear - microtia or totally absent ear - anotia), preauricular appendages, auricular fistulae, maxillary or mandibular hypoplasia, vertebral deformities, hemifacial microsomia and vertebral anomalies. Dermoids can be associated with ocular abnormalities including colobomata of the eyelids, Duane retraction syndrome and other ocular motility disorders, lacrimal anomalies, scleral and corneal staphylomata, aniridia, and microphthalmia. Variant of the syndrome like a fibroepithelial polyp attached to limbal dermoid has also been described⁵. Unilateral morning glory syndrome has been found in a patient with multiple limbal dermoids⁶. Associations like SCALP^{7,8} and Nager syndrome⁹ are reported in the literature. A new grading system keeping in view area of cornea and conjunctiva involved as well as surface shape has been proposed¹⁰. Reviewing Pakistani literature on the subject found a few case reports and small studies¹¹⁻¹⁵. This study was carried out to analyze our experience regarding the clinical presentation and results of simple excision of limbal dermoids type one in Pakistani population.

MATERIAL AND METHODS

A total of 15 epibulbar dermoids presented in oculo-plastic and pediatric ophthalmology division of Mughal eye hospital Lahore and were surgically managed from 1st June 2016 to 30 Dec 2017. This study was approved by the Ethics Committee of Mughal Eye Hospital and followed the tenets of the Declaration of Helsinki. Written informed consent was taken from all patients. All the patients presenting with Type one limbal dermoid (i.e. present at the limbus) who were concerned about cosmetic appearance and were willing for surgical removal were included in the study. One case of dermoid which was involving the entire cornea was excluded. Informed consent was taken from all the participants. All the excisions were done by the first author. Adults were operated under local anesthesia and children were operated under general anesthesia. After excision with blade and scissors, conjunctiva was stitched in 6 (40 %) cases when lesion was affecting significant part of conjunctiva. All the operated cases were reviewed in

outpatient department on 1st post-operative day, every week for three weeks and then every month for 4 months. Follow up ranged from 3 weeks to 4 months (Mean= 6 ± 3.5 weeks).

RESULTS

Age ranged from 4 years to 60 years (Mean 18+ 13.48). 3 (20%) out of 15 patients presented in 1st decade of life, 8 (53%) were in second, 3 (20%) in third decade and one (6.7%) patient in 6th decade of life. All patients presented due to cosmetic concerns though 12 cases (80 %) had visual deterioration.

Male to female ratio was 6:9. All the patients had unilateral epibulbar dermoids. In our study all the patients presented with epibulbar dermoid in inferior temporal quadrant however in 3 patients (20%) lesion was more towards inferior aspect of limbus. Most (9 cases - 60%) were present at limbus equally involving cornea and sclera. However, 3 cases (20%) extended more on the scleral side while 3 cases (20%) were predominantly on the corneal side. Most (12 cases-80%) were round and 3 cases (20%) tapering. 5 (33%) had Goldenhar Syndrome (Table 1). 4 (27%) patients

Table 1: Systemic associations of Limbal Dermoid (Total No. of patients =15).

Ocular & Systemic associations	5 (33%)
Preauricular skin tags	4 (27%)
Ear deformity	3 (20%)
Maxillary Hypoplasia	1 (7%)
Divergent Squint	1 (7%)

had preauricular tags. Pre auricular tags were on the same side as the lesion in 2 patients, on opposite side in 1 patient and bilateral in one patient. One (7%) patient had maxillary hypoplasia. She also had divergent squint. Postoperatively (Table 2) one patient (7%) had extensive formation of granulation tissue while one patient (7%) had corneal thinning and his post-operative steroids were stopped immediately.

Table 2: Post operative complications.

Post operative complications	2 (14%)
Corneal thinning	1 (7%)
Extensive formation of granulation tissue	1 (7%)

There was variable yellowish to brownish pigmentation of the lesion in 12 cases (80%). Five

patients (33%) out of 15 had more marked superficial pigmentation of epibulbar dermoid. Three (20%) patients had ear deformity (hypoplastic ear - microtia). After excision cornea and conjunctiva healed within 5-7 days, generally with some scarring and imperfect corneal transparency; however, the appearance was considerably improved.

DISCUSSION

Limbal dermoids belong to benign congenital tumors containing choristomatous tissue i.e. normal tissue derived from germ cells layers, which is foreign for that site. There is no racial predisposition and males and females are equally affected. In our study male to female ratio was 6:9. Most common site of presentation of limbal dermoid is inferior temporal quadrant of the corneal limbus. Most of limbal dermoids were equally involving corneal and scleral sides of limbus while in a few patients lesion was more on corneal side or scleral side. Epibulbar dermoids are dome shaped, with or without keratinized surface. Hair follicles and cilia are usually visible. They are fleshy and can have fine superficial vessels. They usually are not malignant. Multifactorial pattern of inheritance is well-recognized in limbal dermoids associated with ocular and systemic findings such as Goldenhar syndrome.

In our study, patients presented at different age groups. The late presentation in our cases was probably due to socio economic reasons as poor patients could not afford early treatment. Fourteen (93%) out of 15 patients presented with superficial epibulbar dermoids while one patient had deep corneal stromal involvement.

Management of limbal dermoids may be conservative with artificial lubricants and epilation of offending hair if there is foreign body sensation. Surgical removal of the lesion can be done in case of cosmetic disfigurement or if it is causing visual disturbance. Surgical treatment is indicated only when there is requirement for improving the patient's vision or cosmetic appearance. Surgical removal of the mass which is above the surface of sclera or cornea is the preferred method. It is unnecessary to completely remove the deeper lesion as inadvertent entry inside eyeball is high in case of repeated attempts for complete excision of the lesion. The exposed sclera is covered with the help of undermining surrounding conjunctiva and suturing it over exposed surface. In case of removal of most thickness of cornea or sclera, a

patch graft is done to restore thickness of the wall of eyeball. Amniotic membrane may be stitched in a single or multiple layers at the site if there is risk of perforation. The amniotic membrane is sutured to underlying sclera or fibrin-glue adhesive is used to secure the grafted tissue^{16,17}. Placement of a processed pericardial graft to cover exposed surface after excision has also been tried¹⁸. In all of our study cases, superficial sclerokeratectomy was done with the help of blade and scissors for excision of epibulbar dermoid. In cases where the epibulbar dermoid was more on scleral side the defects was closed with simple suturing of the conjunctiva. One patient had more deep involvement of corneal stroma with postoperative thinning of the cornea. His postoperative steroids were stopped immediately. One should remain vigilant and should have a plan to apply patch if there is impending perforation. One younger patient operated at the age of 4 years had extensive formation of postoperative granulation tissue. Such cases may be confused to have recurrent keloid¹⁹. Limbal stem cell transplantation from the same patient has been found effective²⁰. Sutureless corneoscleral grafts fixed with fibrin glue are becoming more popular²¹. 0.02% Mitomycin C applied for 2 min following the excision has been claimed to prevent occurrence of pseudopterygium following excision²². Tattooing of the cornea and a conjunctival graft of the same patient after simple excision has been claimed to produce better postoperative appearance^{23,24}. Cosmetic concern remains the main indication for the decision to remove limbal dermoids²⁵. Our study has a few limitations which include relatively small number of cases, short follow up (as most patients were satisfied and did not report for follow up) and not using Mitomycin or amniotic membrane so we cannot comment which is a relatively better procedure. Strength of our study is that we have preoperative and postoperative photograph of each patient with all findings. We achieved satisfactory results by simple surgical removal.



Fig. 1: Limbal Dermoids.



Fig. 2: Upper two rows: Goldenhar Syndrome in 5 cases. Upper Left 3 photo, maxillary hypoplasia & divergent squint
 Lower row (Post op): Left 2 photo, Appearance after excision: Central photo, Corneal thinning: 2nd photo from right, Post Conj congestion: Right photo, Post op marked granulation tissue formation.

CONCLUSION

In our study there was yellowish to brownish superficial pigmentation in epibulbar dermoid in most cases, which is not reported earlier to the best of our knowledge. Treatment with excision and superficial sclerokeratectomy without graft gives satisfactory results. No significant visual threatening complication was encountered.

Author's Affiliation

Dr. Khawaja Khalid Shoab
FCPS, FRCS, MCPS HPE
Health Bridge Hospital, Ghazi Road, Near Bhatta Chowk, DHA, Lahore.

Dr. Tariq Shakoor
MCPS, FCPS
Rahbar Medical & Dental College, Lahore

Dr. Muhammad Shahbaz Amin
MCPS, FCPS
Lahore Medical & Dental College, Lahore.

Role of Authors

Dr. Khawaja Khalid Shoab
Performed Surgery, Review of literature, Collection of Data, Analysis of Data, Writing Manuscript, Critical Proof reading

Dr. Tariq Shakoor
Review of literature, Collection of Data, Analysis of Data, Writing Manuscript, Critical Proof reading

Dr. Muhammad Shahbaz Amin
Review of literature, Collection of Data, Analysis of Data, Writing Manuscript, Critical Proof reading

REFERENCES

1. **Fard AM, Pourafkari L.** Images in clinical medicine. The hairy eyeball – limbal dermoid. *N Engl J Med.* 2013; 368 (1): 64.
2. **Dey R, Dey S.** Images in clinical medicine. Limbal dermoid. *N Engl J Med.* 2011; 364 (6): e9.
3. **Zhu J, Cheng HB, Fan N, Liu CM, Yu WH, Chen XM, Liu XY.** Studies of a pedigree with limbal dermoid cyst. *Int J Ophthalmol.* 2012; 5 (5): 641-3.
4. **Hafidi Z, Daoudi R.** Limbal dermoid in Goldenhar syndrome. *Pan Afr Med J.* 2013; 15: 69.
5. **Seymenoğlu G, Başer E, Tansuğ N, Demireli P.** An unusual association of Goldenhar syndrome. *Int Ophthalmol.* 2013; 33 (1): 91-4.
6. **Lowry EA, de Alba Campomanes AG.** Unilateral Morning Glory Disc Anomaly With Ipsilateral Limbal Dermoids. *J Pediatr Ophthalmol Strabismus.* 2014; 51 Online: e37-9.
7. **Lam J, Dohil MA, Eichenfield LF, Cunningham BB.** SCALP syndrome: sebaceous nevus syndrome, CNS malformations, aplasia cutis congenita, limbal dermoid, and pigmented nevus (giant congenital melanocytic nevus) with neurocutaneous melanosis: a distinct syndromic entity. *J Am Acad Dermatol.* 2008; 58 (5): 884-8.
8. **Hsieh CW, Wu YH, Lin SP, Peng CC, Ho CS.** Sebaceous nevus syndrome, central nervous system malformations, aplasia cutis congenita, limbal dermoid, and pigmented nevus syndrome. *Pediatr Dermatol.* 2012; 29 (3): 365-7.
9. **Malik R, Goel S, Aggarwal S.** Limbal dermoid in Nager acrofacial dysostosis: a rare case report. *Indian J Ophthalmol.* 2014; 62 (3): 339-41.
10. **Zhong J, Deng Y, Zhang P, Li S, Huang H, Wang B, Zhang H, Peng L, Yang R, Xu J, Yuan J.** New Grading System for Limbal Dermoid: A Retrospective Analysis of 261 Cases over a 10-Year Period. *Cornea,* 2018; 37 (1): 66-71.
11. **Moin M, Din I, Nazeer A.** Ocular and Periocular dermoid cysts; a clinicopathological study. *Biomedica* 2005; 21 (2): 113-6.
12. **Kumkum G, Manish J, Prashant K G, Bhawana R.** Goldenhar syndrome: airway and anesthetic management – a case report. *J Pak Med Students* 2011; 1 (2): 56-9.
13. **Nadeem S, Raza A.** Presentation of ocular and orbital dermoid cysts at Holy Family Hospital Rawalpindi. *Pak J Ophthalmol* 2012; 28 (2): 95-8.
14. **Hayat K, Shahzad M, Lin Z, Miao L, Xue S.** Case Report Surgical Therapy Of Unusual Congenital Corneal Fibroma. *J Ayub Med Coll Abbottabad,* 2010; 22 (1): 180.
15. **Kausar A, Zafar SN, Altaf S, Khan A.** Ophthalmic manifestations of linear nevus sebaceous/organoid nevus syndrome. *J Coll Physicians Surg Pak.* 2015; 25 (3): 220-2.
16. **Pirouzian A, Ly H, Holz H, Sudesh RS, Chuck RS.** Fibrin-glue assisted multilayered amniotic membrane transplantation in surgical management of pediatric corneal limbal dermoid: a novel approach. *Graefes Arch Clin Exp Ophthalmol.* 2011; 249 (2): 261-5.
17. **Pirouzian A.** Management of pediatric corneal limbal dermoids. *Clin Ophthalmol.* 2013; 7: 607-14.
18. **Lazzaro DR, Coe R.** Repair of limbal dermoid with excision and placement of a circumlimbal pericardial graft. *Eye Contact Lens,* 2010; 36 (4): 228-9.
19. **Gaviria JG, Johnson DA, Scribbick F 3rd.** Corneal keloid mimicking a recurrent limbal dermoid. *J Pediatr Ophthalmol Strabismus.* 2005; 42 (3): 189-90.
20. **Hong S, Kim EJ, Seong GJ, Seo KY.** Limbal Stem Cell Transplantation for Limbal Dermoid. *Ophthalmic Surg*

- Lasers Imaging, 2010; 9: 1-2.
21. **Zhou AX, Ambati BK.** Sutureless Lamellar Corneoscleral Patch Graft with Fibrin Sealant for Limbal Dermoid Removal. *J Pediatr Ophthalmol Strabismus*. 2016 Jun. 3; 53 Online: e22-5. Doi: 10.3928/01913913-20160509-03.
 22. **Lang SJ, Böhringer D, Reinhard T.** Surgical management of corneal limbal dermoids: retrospective study of different techniques and use of Mitomycin C. *Eye (Lond)*. 2014; 28 (7): 857-62.
 23. **Jeong J, Song YJ, Jung SI, Kwon JW.** New surgical approach for limbal dermoids in children: simple excision, corneal tattooing, and sutureless limboconjunctival autograft. *Cornea*, 2015; 34 (6): 720-3.
 24. **Cha DM, Shin KH, Kim KH, Kwon JW.** Simple keratectomy and corneal tattooing for limbal dermoids: results of a 3-year study. *Int J Ophthalmol*. 2013; 6 (4): 463-6.
 25. **Matsuo T.** Clinical decision upon resection or observation of ocular surface dermoid lesions with the visual axis unaffected in pediatric patients. *Springerplus*, 2015; 4: 534.