Management of limbal dermoid

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Abstract

Epibulbar dermoids are benign, congenital choristomas that may vary in size, location and depth causing variable symptoms like astigmatism, amblyopia or total loss of vision. Their management presents with various challenges because of varied presentation and cosmetic reasons. This article summarizes the various surgical and medical management options for different grades of limbal dermoids along with a case report of a young female who presented with a limbal dermoid with mild astigmatism, which was corrected with glasses.

Keywords: Dermoid, limbal, choristoma.

Introduction

Choristoma is a benign congenital tumor that consists of histologically normal tissue that is derived from germ layers foreign to that anatomic location. Limbal dermoid is a form of epibulbar choristoma constituting collagenous connective tissue with a sebaceous component. They can be unilateral or bilateral, single or multiple, and mostly located on bulbar conjunctiva, cornea or limbus. Amongst limbal dermoids, the most common location is inferotemporal. It is an embryological anomaly occurring at 5-10 weeks gestation in which there is metaplastic transformation of mesoblast between surface ectoderm and rim of the optic nerve. Epibulbar dermoids are congenital lesions with connective tissue hyperplasia with sebaceous component. Histopathological examination shows sebaceous gland acini and hair follicles inside a well-defined nodule of fibrous connective tissue. Limbal dermoid can be isolated finding or may be associated with other ocular findings like aniridia, aphakia, microphthalmia, scleral staphyloma or corneal staphyloma. It can also be associated with some syndromes like Nager acrofacial dysostosis or Goldenhar syndrome which is now named Goldenhar-Gorlin syndrome after including vertebral anomalies. Limbal dermoids are classified into three grades anatomically as described below:

- Grade 1- superficial lesions measuring less than 5 mm and localized to limbus
- Grade 2- lesion covering most of cornea (more than 5 mm) and extending deep into stroma but not involving Descemet's membrane
- Grade 3- lesion covering whole cornea and extending through histological structures between anterior surface of eyeball and pigmented epithelium of iris.

The clinical features may be cosmetic disfigurement or diminution of vision due to astigmatism depending upon the size of dermoid. Management includes medical management and surgical correction.

Case Report

A 17-year-old female presented to our OPD with an asymptomatic lesion in her right eye which was present since birth. She was referred from Medicine department for eye opinion regarding the lesion. She had normal facial features and no skeletal anomalies were noted. Audiometry was done to rule out any hearing deficit, and was found out to be within normal limits. Detailed examination of eye showed nasal limbal dermoid in right eye (Figure 1). Pupil was eccentric and reacting to light.

Fig. 1 : Right eye showing limbal dermoid
Vision (unaided) in right eye was 6/9 and left eye was
6/6. Refraction corrected the vision in right eye to 6/6 with addition of cylindrical lens. Slit lamp examination showed the dimensions of dermoid to be 3mm x 3mm and depth extending to stroma classifying it as grade 2 dermoid. The lesion also showed 2 hair strands projecting out from it. Anterior chamber was of normal depth and other structures also showed no deviation from normal anatomy. (Figure 2) Fundus examination showed no abnormality.

Fig. 2 : Right eye showing normal fundus

Detailed examination of the other eye turned out to be completely normal. Patient was advised removal of lesion under local anaesthesia for cosmetic reasons but she refused for any intervention and didn't come for follow up.

Medical Management.
Management in literature mostly says to “leave these lesions alone”. Medical management is recommended in grade 1 dermoids with up to 1 Dioptre astigmatism. Patients who are managed conservatively with spectacles should be kept on regular follow-up and compliance should be ensured. Regular visits should be planned every 2-3 months. Visual acuity, refraction, size of lesion using digital photography should be monitored at every visit. Possibility of amblyopia should also be kept in mind in pediatric age group. On any visit, if any of above parameters are affected or patient is not compliant, surgery should be considered.

Surgical Management
Dermoids are benign lesions, but they might require surgical excision in many cases. Enlarging lesions can cause astigmatism, disturbance of tear film leading to irritation and frequent rubbing of eyes.

Surgical correction should be considered in conditions described below

| Table 1 |
|-----------------|------------------|
| 1               | Chronic eye rubbing due to irritation and recurrent conjunctivitis |
| 2               | Amblyopia unresponsive to medical management |
| 3               | Progressive dellen, with corneal surface decompensation |
| 4               | Growth encroaching into pupillary area or optical zone |
| 5               | Irregular astigmatism |
| 6               | Inadequate lid closure |
| 7               | Cosmetic indication |

For dermoids greater than grade 2, surgical management is indicated. Surgeries vary from simple excision to keratoplasty. Simple keratectomy is advised for small and superficial lesions, with major disadvantage being the residual astigmatism.

Keratoplasty can be lamellar or penetrating, with only central or full curvature graft. The advantage of these procedures include correction of astigmatism as well as improved cosmesis while the disadvantages being re-epithelization, interface neovascularisation, steroid-induced glaucoma and graft rejection.

Newer techniques include corneal- limbal scleral donor graft transplantation and surgical resection followed by amniotic membrane transplantation. Multilayered amniotic membrane graft is considered better for volumetric filling of the defect created by excision. Some studies have proven autologous limbal stem cell transplantation as a good option to compensate for the defect created due to excision of large dermoid. Recent advances state that use of fibrin glue to transplant amniotic membrane has reduced chances of graft rejection. Ongoing studies suggest the future management of limbal dermoid with pericardial patch graft as reconstructing alternative along with amniotic membrane multilayered graft.

The best surgical option for every case can vary according to various factors and it is summarized in table 2.

| Table 2 : Recommendations for surgical removal of ocular dermoids |
|-----------------|------------------|
| Grade of ocular dermoid | Recommended techniques |
| Grade 1 (≤50µm thickness and ≤1mm diameter) | Simple excision |
| Grade 1 (>50µm thickness and <1 mm diameter) | Keratectomy + AMT + ALSCA |
| Grade 2 and deeper grade 1 | Keratectomy + AMT + ALSCA + PPG vs anterior or deep anterior lamellar keratoplasty + AMT |
| Grade 3 | Total anterior segment reconstruction |
Abbreviations: AMT- amniotic membrane transplant, ALSCA- autologous limbal stem cell allograft, PPG- pericardial patch graft

References

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