Surgical Outcomes of Epibulbar Dermoids

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ABSTRACT

Purpose: To evaluate results of lamellar keratoplasty in limbal dermoid.

Methods: The ocular records were reviewed of 155 consecutive eyes with solid epibulbar dermoids that underwent lamellar keratoplasty at Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences from 1977 to 1998. According to the size and location, the dermoids were managed surgically either by sectoral, annular, or central lamellar keratoplasty.

Results: All but 16 eyes improved cosmetically; while all the patients showed reduction in astigmatism, 116 eyes improved functionally.

Conclusion: Excision with lamellar keratoplasty appears to be an effective means of management for extensive limbal dermoid. To avoid development of amblyopia, surgery at an early age is preferred.


INTRODUCTION

Choristomas represent the most common epibulbar lesions in the pediatric age group. Epibulbar or limbal dermoids are the most frequently encountered ocular benign tumor. Generally, these are superficially placed and do not involve the deeper layers of the cornea or sclera. Lamellar or penetrating keratoplasty has been advocated, depending on the site and depth of the involvements. Since most of the lesions occur in children, keratoplasties have guarded prognosis because of accompanying amblyopia and high incidence of vascularization of the grafts.

We, herein, report the results of analysis of the excision of limbal dermoids, followed by lamellar keratoplasty in 155 eyes of 150 patients over the past 22 years.

MATERIALS AND METHODS

Case records of 155 eyes of 150 patients with limbal dermoids who underwent surgical excision with lamellar keratoplasty at Dr. Rajendra Prasad Centre for Ophthalmic Sciences, New Delhi, over a 22-year period (1977 through 1998), were retrospectively analyzed (Figure 1). Patients who had a follow up at least 1 year following surgery were included in the study.

In addition to the detailed ocular and general histories, the rate of growth of the dermoid at puberty of each patient was reviewed. Findings of slit lamp biomicroscopy to determine the size, site, extent, and depth of the corneal involvement and gonioscopy (whenever available) were analyzed.
Using the classification followed at our center, all lesions had been graded depending on the depth and extent of corneal involvement (Tables 1-3, Figure 2).

The surgical procedure included the excision of the lesion followed by depth-dependent lamellar keratoplasty. The defect was covered by a size and depth-matched fresh eyes (97), MK preserved eyes (48), glycerine preserved eyes (10), and donor corneal tissue (depending on availability) with 10-0 monofilament-interrupted sutures. Postoperatively, all patients received topical diluted 0.1% Betamethasone 1:1 and artificial tears at 2 hour intervals, that were gradually tapered. The patients were followed up daily for 1 week, twice a week the following week, monthly for 3 consecutive months, and at the end of 1 year.

The follow-up records of these patients were further evaluated for noting the best corrected visual acuity at 3 months and following conventional amblyopia therapy. Keratometry (Baush and Lomb keratometre) finding was recorded both preoperatively and after a follow up of 1 year.

Of 77 eyes diagnosed with amblyopia, cycloplegic refraction was done in all eyes, regardless of patient age. The patients were divided into 3 groups based on their ages (Table 4). Conventional occlusion was given to the Group I children by occluding the normal eye with cotton pad and 3M micropore tape. The schedule was age-dependent, 1:1, 2:1, 3:1, and 6:1 for 1, 2, and 3 to 6 years, respectively. Between 6 to 10 years, 6:1 ratio was followed to avoid occlusion amblyopia. The occlusion was weaned off slowly over 6 months to 2 years by initially switching to 1:1 occlusion and to partial occlusion using multiple layers of cello tape over the glasses in the normal eye. The layers were reduced on follow ups if no deterioration of vision was noted in the amblyopic eye. All of these patients were followed up until their 10th birthday. In Group II, a

<table>
<thead>
<tr>
<th>TABLE 1</th>
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<tbody>
<tr>
<td><strong>GRADES OF LIMBAL DERMOID</strong></td>
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</table>

<table>
<thead>
<tr>
<th>Grade</th>
<th>Depth of Involvement %</th>
<th>Extent of Corneal Involvement From Limbus (mm)</th>
<th>Surgery to Be Planned</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>&lt;50</td>
<td>&lt;3</td>
<td>Excision (Ex)</td>
<td>Excellent</td>
</tr>
<tr>
<td>II</td>
<td>&lt;50</td>
<td>3 to 5</td>
<td>Ex</td>
<td>Good</td>
</tr>
<tr>
<td>III</td>
<td>&gt;50</td>
<td>3 to 5</td>
<td>Ex, LK*</td>
<td>Fair</td>
</tr>
<tr>
<td>IV</td>
<td>&gt;50</td>
<td>5.1 to 7</td>
<td>Ex, Ex+LK</td>
<td>Guarded</td>
</tr>
<tr>
<td>V</td>
<td>&gt;50</td>
<td>&gt;7</td>
<td>Ex+LK</td>
<td>Poor</td>
</tr>
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</table>

*LK = Lamellar Keratoplasty.
TABLE 2
GRADE OF LIMBAL DERMOID VS FUNCTIONAL OUTCOME (N = 155)

<table>
<thead>
<tr>
<th>Grade</th>
<th>Number of Eyes</th>
<th>Initial</th>
<th>Visual Outcome</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>%</td>
<td>I</td>
<td>S</td>
</tr>
<tr>
<td>I</td>
<td>5 (3.2)</td>
<td>5</td>
<td>-</td>
</tr>
<tr>
<td>II</td>
<td>33 (21.3)</td>
<td>28</td>
<td>5</td>
</tr>
<tr>
<td>III</td>
<td>50 (32.2)</td>
<td>16</td>
<td>34</td>
</tr>
<tr>
<td>IV</td>
<td>39 (25.2)</td>
<td>5</td>
<td>32</td>
</tr>
<tr>
<td>V</td>
<td>28 (18.1)</td>
<td>1</td>
<td>25</td>
</tr>
</tbody>
</table>

Abbreviations: I = Improved; S = Status quo; W = Worse

Constants occlusion for 2 weeks was given. If improvement was noted on follow up, the patients were followed up on conventional occlusion of 6:1. Four patients had eccentric fixation. They were given inverse occlusion by occluding the abnormal eye followed by pleoptics. Group III patients were given a bolus of constant occlusion for 4 weeks, also followed by pleoptics.

RESULTS

The age of the 150 patients (79 males, 71 females) ranged from 3 months to 26 years (Figure 1). In all patients (145 unilateral and 5 bilateral), the lesions were seen since birth, while 11 patients gave history of rapid growth at the onset of puberty. Seven eyes showed pure corneal dermoid, while 5 lesions were annular in nature. Of the remaining 143 eyes, 76 showed only one-quadrant involvement, and 67 eyes had more than one quadrant involved.

Five eyes (3.2%) belonged to Grade I, 33 (21.3%) to Grade II, 50 (32.2%) to Grade III, 39 (25.2%) to Grade IV, and 28 (18.1%) eyes belonged to Grade V (Table 2, Figure 2).

Seven eyes had involvement of the angle of the anterior chamber in the form of peripheral anterior synechiae. The depth of involvement up to Descemet’s membrane was seen in 59 eyes, of which 26 eyes required central subsequent penetrating keratoplasty within 2 to 5 years. The other 33 eyes had peripheral corneal opacity with sparing of the pupil. The best corrected visual acuity of 6/18 or better was seen in 49 eyes 3 months postoperatively, compared to 13 eyes preoperatively (Figure 3). Of 106 eyes, where visual acuity was <6/18 at the end of 3 months, 70 were due to recipient bed/graft opacity, and in 52 eyes the poor vision was due to dense amblyopia in the presence of the clear graft. Another 15 eyes also had visual acuity 6/18-6/12 due to amblyopia. Following amblyopia therapy, there was gain in visual acuity of one line or better in 68 eyes. Three eyes from Group I, 2 from Group II, and 4 from Group III did not show any improvement.

The cornea was absolutely clear in 101 (65%) eyes. Although there was no raised lesions in 54 (35%) eyes, the bed opacity was still persisting (Figure 4).

Astigmatism <3D was achieved in 51 (33%) eyes postoperatively, compared with 88 (57%) preoperatively. Astigmatism that could be assessed preoperatively due to extensive lesion in 20% of eyes was reduced to almost one-half (11%) postoperatively (Figure 5).

None of the eyes had recurrence in the follow-up period of 1 to 18 years. Five eyes developed pseudopterygium and were repaired by recession and repeat keratoplasty, of which 3 improved.

Figure 3: Visual acuity in epibulbar dermoid.
TABLE 3

<table>
<thead>
<tr>
<th>Visual Acuity</th>
<th>Preoperative</th>
<th>3 Months</th>
<th>1 Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>6/6</td>
<td>2(1.3)</td>
<td>2(1.3)</td>
<td>12(7.7)</td>
</tr>
<tr>
<td>6/9</td>
<td>6(3.8)</td>
<td>16(10.3)</td>
<td>29(18.7)</td>
</tr>
<tr>
<td>6/12</td>
<td>3(1.9)</td>
<td>14(9)</td>
<td>15(9.6)</td>
</tr>
<tr>
<td>6/18</td>
<td>2(1.3)</td>
<td>17(10.9)</td>
<td>20(12.9)</td>
</tr>
<tr>
<td>6/24</td>
<td>16(10.3)</td>
<td>10(6.4)</td>
<td>7(4.5)</td>
</tr>
<tr>
<td>6/36</td>
<td>33(21.3)</td>
<td>19(12.2)</td>
<td>4(2.6)</td>
</tr>
<tr>
<td>6/60</td>
<td>25(16.1)</td>
<td>23(14.7)</td>
<td>17(10.9)</td>
</tr>
<tr>
<td>&lt;6/60</td>
<td>68(44)</td>
<td>54(35)</td>
<td>51(33.1)</td>
</tr>
</tbody>
</table>

DISCUSSION

Dermoid of the limbus and cornea are solid tumors and are classified as hamartia because they contain cellular elements not normally present in that location, such as ectodermal derivatives (hair follicles), as well as sebaceous and sweat glands embedded in connective tissue and covered by squamous epithelium. Dermoid cysts and dermolipectes are also hamartia, but they occur in the ocular adnexa, not on the globe itself.10

Limbal dermoids have been classified under 3 categories by the extent of their involvement: 1) Small—straddling at limbus up to 5 mm in size; 2) Large—covering entire corneal surface but not progressing beyond Descemet's membrane; and 3) Extensive—replaces cornea, anterior chamber, and iris stroma and is lined posteriorly pigment epithelium of the iris. However, in the present study, we used our own grading.

The extensive category according to this classification, is more of a histopathological diagnosis. Since all the specimens were not subjected to histopathological examination, we graded the same based on clinical findings. We believe this classification will be helpful from both the therapeutic and prognostic point of view. The grading will also enable the corneal surgeon to decide about which surgical procedure to follow.

Management of epibulbar tumors is only by surgery.1,4-8,13 The indications of surgery are for cosmetic reasons, irritative symptoms because of the growth, hair on the growth, poor vision either due to encroachment of the cornea (by the tumor or the lipid infiltration line in front of the mass) or to high astigmatism, and because of induced diplopia.1 Similar, the type of surgery is dependent upon the size, site, depth of involvement, and nature of the growth. Though ultrasound biomicroscopy is beneficial to assess depth of involvement in opaque corneal lesions for facilitating better planning of the surgical approach, we do not have access to the equipment.14,15

Small tumors do not require excision for cosmetic reasons because sometimes the postoperative scar and complicated pseudopterygium produces more cosmetic blemish than the growth itself.16 Furthermore, a simple excision is not sufficient to manage the extensive lesion because keratectomy conducted during excision gives rise to surface irregularity, thus leading to tear fluid abnormalities,17 which, in the long run, can give rise to vascularized corneas. Moreover, there is every chance of formation of pseudopterygium and symblepharon following excision of extensive lesions. Therefore, simple excisions alone, or with keratectomy, are recommended in lesions less than 5 mm in size with superficial involvement.4,18 Moreover, in view of the occurrence of pseudopterygium following excision of limbal dermoid, lamellar keratoplasty is recommended.5

Figure 4.
TABLE 4
AMBLYOPIA (N = 77)

<table>
<thead>
<tr>
<th>Age</th>
<th>Number of Eyes</th>
<th>Therapy Given</th>
<th>Gain</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 to 10</td>
<td>51</td>
<td>51</td>
<td>48</td>
</tr>
<tr>
<td>11 to 20</td>
<td>15</td>
<td>15</td>
<td>14</td>
</tr>
<tr>
<td>&gt;20</td>
<td>11</td>
<td>11</td>
<td>8</td>
</tr>
</tbody>
</table>

Sometimes, the dermoids are extensive externally and internally. Though lamellar keratoplasty is not effective in these eyes, no penetrating keratoplasty should be tried as first attempts. In such a situation, a large diameter lamellar keratoplasty should be performed after excising the main part of the growth. Subsequently, a small, central penetrating keratoplasty would be sufficient for restoring the vision.

Our results are in concurrence with Baum et al., who concluded that if the dermoid involves the posterior half of the stroma, the semiopaque area should be left as such, or in other words, an incomplete removal should be done rather than entering the anterior chamber. Simple excision with keratectomy was recommended in cases less than 5 mm in size involvement of superficial stroma. However, in this series, only 5 eyes for whom supple excision was possible belonged in this category.

Total or partial papillary involvement is responsible for amblyopia either because of total occlusion or high astigmatism. In the present series, 65 eyes had total papillary involvement and another 33 eyes had high astigmatism, many of which had poor visual acuity. Moreover, as high as 77 eyes developed amblyopia of which 51 belonged to the 0 to 10 years age group, 15 in the 11 to 20 years age group, and 11 eyes of patients in the more than 20 years age group. Of the 77 eyes having poor vision due to amblyopia either because of papillary area involvement or high astigmatism, 69 eyes gained vision partially or better following conventional amblyopia occlusion therapy. Improvement of vision following amblyopia therapy was excellent in patients under the age of 10 years and only 3 patients failed to respond to therapy. Further, eyes having dense amblyopia responded more poorly to therapy than the eyes with less dense amblyopia where preoperative visual acuity was 6/18 or more.

Another interesting factor observed in this study was male children were operated on at relatively earlier ages than were females. More girls seek ophthalmologist's advice towards later adolescence. Thus, boys developed less amblyopia than the girls overall.

Operative complications were in the form of residual opacity in 54 eyes, pseudopterygium in 2 eyes, and infection in 2 eyes. No recurrence of the tumor in any of the 155 eyes reflects its benign nature and safety of surgical technique.

Astigmatism was reduced remarkably following lamellar keratoplasty. Of the 54 eyes having deeper involvement, 33 were off the visual axis and 26 were occluding vision. These 26 grafts remained opaque for which subsequent penetrating keratoplasty was performed. Thus, the criteria for surgery in such patients are:

- Cosmetic
- Progressive and irritative lesions
- Vision threatened because of astigmatism or papillary involvement
- Fear or amblyopia

The earlier the surgery is performed, the simpler the procedure is and the better cosmetic and functional results are. Seventy-seven eyes had poor vision due to amblyopia, either because of papillary involvement or high astigmatism, of which 51 eyes gained vision following conventional amblyopia occlusion therapy. Preoperative astigmatism was reduced in all eyes. Twenty-six grafts remained opaque either because of central residual opacity or otherwise occluding the vision for which subsequent penetrating keratoplasty was performed.

**CONCLUSION**

In conclusion, our series is the largest yet in the world literature and shows that excision of limbal dermoid with lamellar keratoplasty in eyes with deep and large lesion still gives encouraging results.

![Image: Figure 5: Astigmatism in epibulbar dermoid](image-url)
both from an anatomical and a functional point of view. However, it was evident that the simpler the procedure, the better the cosmetic and functional results. Finally, we believe the criteria for surgery, excision plus lamellar keratoplasty, should be for cosmetic reasons in progressive and irritative lesions, vision threatening due to astigmatism or papillary area encroachment, and for fear of amblyopia. Moreover, on the basis of our new classification, the prognostication can be attempted.

REFERENCES