Periocular Skin Malignancies

Jeffrey Buyten, MD
Faculty Advisor: Vicente Resto, MD, PhD
The University of Texas Medical Branch
Department of Otolaryngology
Grand Rounds Presentation
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Outline

- Anatomy
- Periocular BCCA and SCCA
- Treatment options
  - Mohs
  - Non-Mohs excision
  - Radiation
- Orbital invasion
- Orbital exenteration
- Reconstruction
Periocular Definition

- Brow – superior
- Infraorbital rim - inferior
- Nose - medial
- Lateral orbital rim - lateral
Anatomy

- **Eyelid functions**
  - Protect eye (light, injury, desiccation)
  - Tear production and distribution

- **Anterior lamella = skin + obicularis**
- **Posterior lamella = tarsus + conjunctiva**

- **Extremely thin skin (upper > lower)**

- **Skin**
  - Little subcutaneous fat
  - Adherent over the tarsus (levator aponeurosis)
- Horizontal length – 30 mm
- Palpebral fissure – 10 mm

- Margin reflex distance
  - Upper lid – 4 to 5 mm  
    (slightly below limbus)
  - Lower lid – 5 mm  
    (at the lower limbus)

*Note palpebral aperture measurement is the same for examples A and D.*

Fig. 2-5 The margin reflex distance.
Tarsus

- Dense, fibrous tissue
- Contour and skeleton
- Contains meibomian glands
- Length – 25 mm
- Thickness – 1 mm
- Height
  - Upper plate – 10 mm
  - Lower plate – 4 mm
Orbital Septum

- Fascial barrier
- Deep to posterior orbicularis fascia
- Anterior extent of orbit
- Posterior extent of eyelid
Canthal Tendons

- Whitnall's tubercle
- Medial canthal tendon posterior limb
- Posterior lacrimal crest
- Lacrimal sac
- Medial canthal tendon anterior limb
- Anterior lacrimal crest
Canthal Tendons
Fig. 9-1  The anatomy of lacrimal system.
Basal Cell Carcinoma (BCCA)

- 80% on head and neck
- 15% on trunk and arms
- Accounts for 14% of lid lesions
- 80-95% of lid & canthal malignancies
- Met rate 0.0028 – 0.55%

BCCA Incidence

- **Australia**
  - Up to 1772/100 000 (men)
  - Up to 1610/100 000 (women)

- **North America**
  - 300/100,000

- **Rates increasing up to 10%/year**

Risk Factors

- Sun exposure
- Fair skin
- Solid organ transplant
- Immunodeficiency
- Genetic predisposition
  - Basal Cell Nevus syndrome
  - Xeroderma Pigmentosum
  - Albinism
  - Bazex Syndrome

Clinical Features

- Up to 60% pts with one BCC have another on the face
- 20-40% missed clinically
- Destructive lesions (rodent ulcer)
- Pain is uncommon

Natural History

- Doubles in size q 6 months
- Lid BCCA grows laterally
  - Tarsal plate acts as a barrier
- Untreated lesions may travel via periosteum into orbit and cranium

Distribution

- Lower lid - 43%
- Medial canthus - 26%
- Upper lid - 12%
- Lateral canthus - 8%

Pathology

- BCCA - proliferation of basaloid cells from epidermis and invading the dermis

- Palisading of peripheral tumor cells

- Retraction artifact – clear space noted b/w tumor nodules and adjacent stroma

Morphological Classification

- Nodular (50%)
  - Pearly papule
  - Telangectasia
  - Rolled border

- Microscopic
  - Retraction artifact present
  - 15% micronodular (subclinical extension)

- Linear variant
  - Periocular predilection
  - May be more aggressive

Morphological Classification

- Superficial (15%)
  - Monoclonal budding tumor nests attached to underside of epidermis
- Scaly erythematous patch / plaque
  - Mimics psoriasis, discoid eczema, in-situ SCCA or Bowen dz
- Trunk > H&N
- Contains melanin
  - Brown, blue or black

Morphological Classification

- **Infiltrative (10-20%)**, 
  - Irregular groups of tumor cells with a spiky appearance 
  - Palisading and retraction artifact absent

- **Morpheic variant (5%)**
  - Indurated, scar-like plaque w/indistinct margins 
  - Irregular islands & cords of tumor cells 
  - Infiltrate into dense, sclerotic stroma 
  - Increased recurrence risk

- **Mixed (10-15%)**
  - Combo of subtypes

High Risk Lesions

- Incomplete excision
- Recurrent
- Prior non-surgical therapy
- Size > 2 cm
- H-zone
- Medial canthus
  - Highest risk of recurrence
- Poorly defined margins
- Infiltrative / morpheic / micronodular
- Perineural invasion
- Immunosuppression

Table 7. Histological Subtype and Recurrence

<table>
<thead>
<tr>
<th>Main Histological Subtypes</th>
<th>No. of Cases of Primary BCC (840)</th>
<th>No. of Cases of Recurrent BCC (399)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infiltrating</td>
<td>275 (33%)</td>
<td>156 (39%)</td>
</tr>
<tr>
<td>Nodulocystic</td>
<td>379 (45%)*</td>
<td>110 (28%)</td>
</tr>
<tr>
<td>Superficial</td>
<td>85 (10%)</td>
<td>84 (21%)*</td>
</tr>
</tbody>
</table>

BCC = basal cell carcinoma.

*P<0.0001.

Table 4. Recurrence after Mohs’ Micrographic Surgery (MMS) at 5-Year Follow-up

<table>
<thead>
<tr>
<th>Post-Mohs’ Recurrence</th>
<th>Site</th>
<th>Year of MMS</th>
<th>No. of Prior Recurrences</th>
<th>Histological Subtype</th>
<th>Prior Treatments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Medial canthus</td>
<td>1993</td>
<td>2</td>
<td>Infiltrating</td>
<td>Surgical excision, cryotherapy</td>
</tr>
<tr>
<td>2</td>
<td>Medial canthus</td>
<td>1994</td>
<td>1</td>
<td>Infiltrating</td>
<td>Surgical excision</td>
</tr>
<tr>
<td>3</td>
<td>Medial canthus</td>
<td>1994</td>
<td>1</td>
<td>Infiltrating</td>
<td>Surgical excision</td>
</tr>
<tr>
<td>4</td>
<td>Medial canthus</td>
<td>1994</td>
<td>1</td>
<td>Infiltrating</td>
<td>Surgical excision</td>
</tr>
<tr>
<td>5</td>
<td>Lower eyelid</td>
<td>1995</td>
<td>1</td>
<td>Superficial</td>
<td>Surgical excision, MMS</td>
</tr>
<tr>
<td>6</td>
<td>Medial canthus</td>
<td>1995</td>
<td>2</td>
<td>Superficial</td>
<td>Surgical excision, MMS</td>
</tr>
<tr>
<td>7</td>
<td>Lower eyelid</td>
<td>1996</td>
<td>3</td>
<td>Infiltrating</td>
<td>Two cryotherapies, MMS</td>
</tr>
</tbody>
</table>

Perineural Invasion

- Histologic perineural invasion in 1-3%

- More common in morpheic tumors
  - Risk factor for orbital invasion
BCCA Recurrence Rates

- 66% of recurrences occur w/in 3 yrs
- 18% of recurrences occur b/w 5 – 10 yrs

Traditional surgical excision recurrence rates
- 23.4 - 34.8%

Non-Mohs 5 year recurrence rate
- En face frozen section technique
  - Primary 2.1%
  - Recurrent 4.4%

Mohs 5 year recurrence rate
- Primary tumors 1-2%
- Recurrent tumors 5.6-7.8%

p53 and Bcl-2

- p53-positive samples
  - Sclerosing subtype
  - Correlated with aggressive behavior

- Bcl-2 expression
  - 21 low-risk BCCs
  - 10 sclerosing BCCs
  - Associated with slow tumor growth

Shh and Gli1

- Sonic hedgehog signal activates Gli1 transcription factors
- Deregulation of Shh-Gli pathway implicated in familial and sporadic BCCA
- Nearly all BCCA lesions express Gli1
- SCCA does not express Gli1

Squamous Cell Carcinoma (SCCA)

- 5 - 10% of periocular malignancies
- Presents as keratinized plaque
- Erosions or ulcers
- More rapid growth

5-year metastatic rates
- 5 - 45%
- Overall < 10%

Figure 2  A large eyelid squamous cell carcinoma that invaded the orbit in a patient with epidermodyplasia verruciformis. Informed patient consent was obtained for publication of this figure.

1. Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
SCCA Incidence

- **Australia**
  - 600/100000 men
  - 298/100000 women

- **USA**
  - 40-158/100000 men
  - 13-56/100000 women

Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
SCCA Histology

- Well diff – 50.7%
- Moderately diff – 36.6%
  - More subclinical extension
  - Larger difference in tumor and defect size ($P = 0.0021$)
- Poorly diff – 5.6%

Table 5. Periocular Site of Squamous Cell Carcinoma (SCC) and Histology

<table>
<thead>
<tr>
<th>Site of SCC (%)</th>
<th>Lower Eyelid</th>
<th>Upper Eyelid</th>
<th>Medial Canthus</th>
<th>Primary SCC (%)</th>
<th>Recurrent SCC (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Overall (79)</td>
<td>54 (68.4)</td>
<td>6 (7.6)</td>
<td>19 (24)</td>
<td>55</td>
<td>19</td>
</tr>
<tr>
<td>SCC histology</td>
<td></td>
<td></td>
<td></td>
<td>Primary SCC (%)</td>
<td>Recurrent SCC (%)</td>
</tr>
<tr>
<td>Acantholytic (5)</td>
<td>4 (80)</td>
<td>0</td>
<td>1 (20)</td>
<td>4 (80)</td>
<td>1 (20)</td>
</tr>
<tr>
<td>Poorly differen</td>
<td>4 (100)</td>
<td>0</td>
<td>0</td>
<td>1 (25)</td>
<td>3 (75)*</td>
</tr>
<tr>
<td>Moderately differ</td>
<td>16 (61)</td>
<td>2 (8)</td>
<td>8 (31)</td>
<td>21 (81)</td>
<td>5 (19)</td>
</tr>
<tr>
<td>Well differen</td>
<td>27 (75)</td>
<td>3 (8)</td>
<td>6 (17)</td>
<td>26 (72)</td>
<td>10 (28)</td>
</tr>
</tbody>
</table>

No details were available for 8 patients. Periocular site was not associated with any histological subtype ($P = 0.0799$). There was no association between prior recurrence and gender ($P = 0.2217$), age ($P = 0.2800$), or periocular site ($P = 0.4651$).

*P = 0.1954.
Perineural Invasion

- Histologic perineural invasion in 2.5 - 14%

- Associated with:
  - Large SCCs (2 cm)
  - Head and neck tumors
  - Prior recurrence
  - Tendency to be poorly differentiated (p = 0.1954)
  - Aggressive tumor behavior

- Poor prognosis.

Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
High Risk Lesions

- Incomplete excision
- Recurrent
- Prior non-surgical therapy
- Lesions on lip, ear, or eyelid
- Size $> 2$ cm
- Poorly differentiated lesions
- Deep lesions
  - $> 4$ mm reported to have 45% metastatic rate
- Perineural invasion
- Scar carcinomas
- Immunocompromised patients

Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
Neck and Parotid Management

- Must evaluate parotid gland and cervical nodal basins in high risk patients and lesions.

- Parotidectomy and neck dissection

- Survival factors
  - Immunosuppression
  - Advanced clinical parotid stage (P3)
  - Pathologic neck node involvement

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O’Brien, CJ et al. The Parotid Gland as a Metastatic Basin for Cutaneous Cancer. ARCH OTOLARYNGOL HEAD NECK SURG/VOL 131, JULY 2005

Table 4. Possible Revised Staging With Separation of Parotid (P) and Neck (N) Disease

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>Single parotid lymph node ≤3 cm in diameter</td>
</tr>
<tr>
<td>P2</td>
<td>Node &gt;3 cm and up to 6 cm in diameter or multiple nodes</td>
</tr>
<tr>
<td>P3</td>
<td>Node &gt;6 cm in diameter or facial nerve or skull base involvement</td>
</tr>
<tr>
<td>N0</td>
<td>Neck disease absent</td>
</tr>
<tr>
<td>N1</td>
<td>Single node ≤3 cm in diameter</td>
</tr>
<tr>
<td>N2</td>
<td>Node &gt;3 cm in diameter or multiple nodes</td>
</tr>
</tbody>
</table>
SCCA Recurrence Rates

- 5-year local recurrence rates (non-Mohs)
  - Vary b/w 3 - 23%

- 5-year recurrence rates (Mohs)
  - Skin 3.1%
  - Lip 2.3%
  - Ear 5.3%
  - Recurrent SCCA 10%

- Study recurrence rate 4%

Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
Excision Margins for SCCA

- Guidelines (Based on MMS studies of 141 tumors)
  - 4-mm margins required to achieve a 95% clearance rate in low-risk SCC
  - 6-mm margins required to clear 95% high risk SCCA
    - > 2 cm
    - Periocular SCCs
    - Histologic grades 2 to 4 (<75% keratinizing cells)
    - Should include subcutaneous fat (30% invade to this level)

Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
Basosquamous Carcinoma (BSC)

- Differentiation of BCCA to SCCA or vice versa
  - Immunostaining has shown true transition zones in lesions

- Collision Tumor
  - Adjacent, separate SCCA and BCCA lesions

- Keratinizing BCCA
  - Variant of nodular BCCA
  - Abrupt keratinization at center of nodule

Basosquamous Carcinoma

- Need to rule out collision tumor and nodular variant.
- If BSC confirmed, then more aggressive treatment is warranted.
  - Recurrence rate of 51.6%
  - More aggressive
  - Met rate 9.7%

Mohs

- Frederick E Mohs - 1941

- Best success rate
  - 5 yr cure rate (1986 periorbital cases)
    - 99% BCCA
    - 98% SCCA

- Confirmed by numerous studies

Mohs Surgery

- Excises tissue in successive layers
- Microscopic evaluation of entire excised surface
- Specimen divided into mapped sections
- Minimizes loss of normal tissue
  - Better reconstruction options
  - Less loss of function
- Ideal for infiltrative tumors

Mohs Surgery

- Follows margin until clear
- Stages = layers of excision
Periocular Mohs Defects

- 264 pts reviewed

- BCCA defects 4.2 - 4.6 x bigger than original tumor
  - Morpheaform → most stages with largest defects
  - Morpheaform defect size 6.1 x larger than original tumor

- Lateral canthus
  - Fewest tumors
  - Largest defects (mean 9.5 cm²)

- SCCA defect size 2.6 x larger than original tumor

## Clinical Tumor and Post Mohs Defect Comparison

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Tumor Area (cm²)</th>
<th>Post-Mohs (cm²)</th>
<th>Δ Size</th>
<th># of Stages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nodular</td>
<td>0.56</td>
<td>2.6</td>
<td>4.6 x</td>
<td>1.8</td>
</tr>
<tr>
<td>Basosquamous</td>
<td>1.5</td>
<td>6.4</td>
<td>4.2 x</td>
<td>2</td>
</tr>
<tr>
<td>Morpheic</td>
<td>1.5</td>
<td>9.2</td>
<td>6.1 x</td>
<td>2.3</td>
</tr>
<tr>
<td>SCCA</td>
<td>0.8</td>
<td>2.1</td>
<td>2.6 x</td>
<td>1.3</td>
</tr>
<tr>
<td>Recurrent</td>
<td>0.7</td>
<td>3.6</td>
<td>5.1 x</td>
<td>1.8</td>
</tr>
</tbody>
</table>

FIG. 2. Combined post-Mohs defect sizes in the periorcular region. Top number is the average defect size. Lower number represents the median defect size.
Surgical Treatments

- Mohs not widely available

- Traditional surgical excision recurrence rates from 23.4 to 34.8%

- Reports of 0% recurrence rate
  - Excision mostly controlled by frozen section study of the margins

# Procedures for Surgical Excision of Basal Cell Carcinoma

<table>
<thead>
<tr>
<th>Technique</th>
<th>Procedure</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Standard excision with postoperative histology</td>
<td>The extent of tumor is marked clinically and excised with a 2–4-mm margin. The defect is repaired and the specimen sent for histology.</td>
<td>Completeness of excision depends on extent of surgical margin (see discussion). There is at least a 30% risk of recurrence with incompletely excised tumors.¹⁸</td>
</tr>
<tr>
<td>Excision with delayed repair</td>
<td>As above, but repair is delayed until the pathology is reported, usually within 24 hours. This allows further excision if the initial surgical margins were not clear of tumor.</td>
<td>May be especially suitable for small, nodular BCC.⁷⁴,⁷⁵</td>
</tr>
<tr>
<td>Excision with frozen section control</td>
<td>The excised specimen is sent immediately for frozen section analysis of tumor margins. Further excision or repair follows depending on the pathology report.</td>
<td>Risk of false-negative results (1–29%).⁷⁶ Using en face sections can minimize false-negative rates (0.2%).⁵³</td>
</tr>
<tr>
<td>Mohs technique</td>
<td>Layered excision (usually 2-mm layers) of the entire lesion is performed. These layers are mapped and embedded so that the entire surgical margin can be examined microscopically. Processing for histology is same as for the frozen section technique.</td>
<td>Lowest risk of recurrence.⁷³ Especially useful for medial canthal, infiltrating, or recurrent BCC. Capital intensive, requires special training.</td>
</tr>
</tbody>
</table>

BCCA Excision & Paraffin Section

- Tumors excised with 4 mm margins
  - Sent for standard paraffin sections

- Primary closure w/o undermining
  - Well demarcated tumors

- Delayed closure (2 – 4 days)
  - Poorly demarcated tumors
  - Large defects requiring more complex repair

BCCA Excision & Paraffin Section

- Complete removal at primary excision
  - 84%

- Repeat excision specimens
  - 53% with no tumor

- 5 year recurrence rate
  - 4.35% (all infiltrative tumours)

BCCA and Overnight Paraffin Section

- 3-mm margin excision
- Rapid (24-hour) paraffin sections
- Re-excision for positive margins
- Reconstruction once margins clear

Recurrence Rate for Overnight Paraffin Sections

- Primary BCC recurrence rate (1/81)
  - 1.23%

- Recurrent BCC recurrence rate (1/7)
  - 12.5%

BCCA and En-face frozens

- Similarities to Mohs
  - Tangential margin examination
  - Quick margin feedback
- Differences from Mohs
  - Surgeon and Pathogist are not the same person
  - Sections examined in vertical slices
- 653 lesions
- 5 year recurrence rate
  - Primary 2.1%
  - Recurrent 4.4%

En Face Frozen Technique

- Pathologist and Surgeon examine pt together in OR. Pathologist present before and during excision.
- Diagram of lesion made
- En face or tangential section of critical margins
  - medial and lateral margins of eyelid
  - adjacent to canaliculi and conjunctival surfaces
  - clinically suspicious margins
- Frozen, sectioned, stained and examined

- Pathologist and Surgeon discuss results in OR
- Re-excision performed if necessary

SCCA and Frozens

- Standard surgical excision
- Frozen en-face section control
  - 36 periocular SCCs
  - 2.8% recurrence at 6 years

Malhorra, R et al. The Australian Mohs Database Periocular Squamous Cell Carcinoma Ophthalmology Volume 111, Number 4, April 2004
SCCA non Mohs excision

- 10 yr retrospective
- 76 pts
- 4 – 5 mm margins
- Primary closure for small defects
- Delayed closure for large defects

SCCA non Mohs excision

- 63 pts → wide surgical excision
- Postop XRT - 20.6%
  - Positive tumour surgical margins
  - Detection of perineural infiltration
- 14 pts → exenteration
- 3 pts → combined bone resection
- 4 pts → sinusectomy

SCCA non Mohs excision

- Group A < 1 cm (n=21)
  - No recurrences
- Group B > 1 cm w/o orbital invasion (n=22)
  - 1 recurrence
- Group C > 1 cm w/orbital invasion (n=33)
  - Recurrence and residual tumor in 16 cases

- Total recurrence rate 22.4%

Radiation Therapy (XRT)

- Poor surgical candidates
- Unresectable tumors
- Full thickness of lid irradiated

Relative contraindications
- Pts under 60 yrs
- Readily excisable tumors
- Tumors over the lacrimal gland

Side Effects

- Erythema
- Skin atrophy
- Subcutaneous fibrosis
- Ulcers
- Epiphora
- Dry eye
- Cataract formation
- Neovascular glaucoma
- Radiation retinopathy
- Radiation optic neuropathy

Superficial XRT for BCCA

- 148 pts (175 lesions)
- 10 yr span (81-91)


| TABLE 4 | Anatomic Distribution for Each Subtype of Basal Cell Carcinoma |
|------------------|------------------|------------------|
| Localization     | Nodular (%)      | Superficial (%)  | Sclerosing (%) |
| Head             | 81.5             | 32.0             | 91.5           |
| Scalp            | 2.9              | 4.0              | 0.0            |
| Forehead/temple  | 17.5             | 12.0             | 12.8           |
| Chin             | 1.9              | 0.0              | 2.1            |
| Cheek            | 4.8              | 4.0              | 4.3            |
| Nose/nasolabial  | 34.0             | 8.0              | 53.1           |
| Eyelid           | 10.7             | 4.0              | 4.3            |
| Ear/periauricular| 8.7              | 0.0              | 12.8           |
| Lip              | 1.0              | 0.0              | 2.1            |
| Trunk            | 13.6             | 56.0             | 6.4            |
| Upper limb       | 1.0              | 0.0              | 2.1            |
| Lower limb       | 3.9              | 8.0              | 0.0            |
| Genitalia        | 0.0              | 4.0              | 0.0            |
Recurrence Rates

- Overall - 15.8%
- Nodular - 8.2%
- Superficial - 16%
- Sclerosing - 27.2%

FIGURE 3. Cumulative 5-year recurrence rates for different basal cell carcinoma subtypes (Kaplan-Meier analysis).

Periocular XRT

- 631 cases BCCA reviewed

- 55 pts treated w/primary xrt
  - Time from xrt to recurrence → 5.3 yrs
  - 7/59 lesions recurred (all from medial canthus)
    - 3/7 → exenteration

- Lesions < 10 mm → 2% recurrence rate
- Lesions > 10 mm → 9.5% recurrence rate

Periocular XRT

- Overall cure rate
  - Primary tumors 98.1%
  - Recurrent tumors 93.6%

Figure 4  Computed tomography of an advanced stage of a recurrent eyelid squamous cell carcinoma that extended into paranasal sinuses and contralateral orbit.
Signs of Orbital Invasion

- Painless mass
- Tumor fixation to bone
- Limited ocular motility
  - Diplopia on extreme gaze (early sign)
- Globe displacement
- Ptosis
- Proptosis - uncommon

Table 2. Signs of Patients with Orbital Invasion from Basal Cell Carcinoma

<table>
<thead>
<tr>
<th>Sign</th>
<th>No. of Patients (N = 56)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visible or palpable mass</td>
<td>56 (100%)</td>
</tr>
<tr>
<td>Mass fixated to orbital bone</td>
<td>20 (35.7%)</td>
</tr>
<tr>
<td>Limitation of ocular motility</td>
<td>17 (30.4%)</td>
</tr>
<tr>
<td>Globe displacement</td>
<td>10 (17.8%)</td>
</tr>
<tr>
<td>No bone fixation and no orbital signs*</td>
<td>20 (35.7%)</td>
</tr>
<tr>
<td>Other signs (epiphora, ptosis, skin fistula)</td>
<td>7 (12.5%)</td>
</tr>
</tbody>
</table>

*Orbital signs: limitation of ocular motility and/or globe displacement. Data not available for 8 patients.
Orbital Invasion

- Can extend along periosteum w/o bony destruction
- Intraocular invasion very rare
- Intracranial spread via superior orbital fissure and cranial foramina

- PNI in 19.3% of patients
  - 91.7% had recurrent tumors
  - Aggressive histologic subtypes

Duration

- Time b/w first detection and invasion of the orbit
  - 2-25 years
  - BCCA - 9.8 yrs
  - SCCA - 1 yr

Frequency

- Reported frequencies of orbital invasion
  - 0.8 to 3.6% BCCA
  - 0.2 to 8.2% SCCA

- 622 pts $\rightarrow$ 2.5%
- 465 patients $\rightarrow$ 3%

Fig. 3  Recurrent postirradiated basal cell carcinoma of medial canthus. Tumour is fixed to and infiltrating bone.
Management of Orbital Invasion

- Imaging
  - CT
  - MRI with contrast – T1

- Extensive orbital invasion present
  - Orbital exenteration followed by XRT

- Anterior orbital involvement / pts with one good eye
  - Local resection with margin control & delayed closure

CT findings

- **Common:**
  - Soft tissue involvement
    - Homogenous, mildly enhancing mass, with irregular borders
  - Bone involvement 20.6%

- **Uncommon:**
  - Rectus muscles infiltration
  - Lacrimal sac extension
  - Ethmoid extension
  - Cribriform plate extension
  - Invasion via superior orbital fissure to involve the dura, cavernous sinus, and cerebral tissue

Treatment

- Exenteration alone
  - 56%
- Exenteration with post op XRT (50–70 Gy)
  - 19%
- Excision alone
  - 9%
- Excision with post op XRT (50–70 Gy)
  - 9%
- Radiotherapy alone (50–70 Gy)
  - 6%

Orbital Exenteration

- 40–50% of exenterations that present to ophthalmologists are required for eyelid or periocular skin tumors

Orbital Exenteration: Absolute Indications

- Involvement of the orbital apex
- Involvement of the extraocular muscles
- Involvement of the bulbar conjunctiva or sclera,
- Lid involvement beyond a reasonable hope for reconstruction
- Nonresectable full thickness invasion through the periorbita into the retrobulbar fat
Orbital Exenteration

- Globe and orbital content removal within the bony socket

Extended Excision

- May involve excision of:
  - Eyelids
  - Bony walls of the orbit
  - Sinuses

- 32 pts
  - Maxillectomy (8)
  - Ethmoidectomy (7)
  - Excision of the frontal sinus (5)
  - Nose (5)

Figure 4  A patient with a basal cell carcinoma following extensive exenteration, rhinectomy, left maxillectomy, ethmoidectomy, and excision of frontal sinus with reconstruction using (a, b) split skin graft and (c, d) orbital prosthesis.
Mortality after Exenteration

- 1 year survival → 93%
- 3 year survival → 67%
- 5 year survival → 57%
- 10 year survival → 37%

Mortality after Exenteration

- **Cause of death:**
  - 54% - orbital tumour
  - 37.5% - unrelated medical conditions
  - 8.3% - distant site malignant processes

- No significance in overall 5 year survival between BCCA and other tumors
- BCCA deaths unrelated to original tumor

Lower Lid Full Thickness Defects

- 0-33%
  - Primary closure
  - Up to 40% w/lid laxity

- 33-50%
  - Tenzel flap

- 50-100%
  - Hughes procedure

Primary Closure

A. Excised Tissue
   - Tarsus
   - Skin and Muscle

B. Placement of marginal sutures

C. [Diagram of suturing process]
Figure 4  Right lower lid surgical defect following BCC excision and 2 month post horizontal direct closure lid reconstruction.
Tenzel Flap

- Semicircle flap based at lateral canthus
- Skin-muscle flap raised
- Cantholysis and canthotomy
- Advance and re-approximate
Hughes Procedure

- Posterior lamella
  - Tarsoconjunctival flap
- Anterior lamella
  - Free skin graft
  - Myocutaneous flap
- 2nd stage
  - Tarsoconjunctival flap takedown
Hughes Procedure
Mustarde Cheek Flap

- Large lower lid lesions
- Single stage
Upper Lid Full Thickness Defects

- Direct closure
- Tenzel flap – inferiorly oriented
- Sliding tarsocconjunctival flap
- Cutler-Beard bridge flap
Sliding Tarsalconjunctival Flap

A

B

C

D

Tarsal-conjunctival flap
Orbital rim
Skin graft
Cutler-Beard Bridge Flap
Cutler-Beard Bridge Flap
Partial thickness defects

- Local advancement / rotational flaps
- FTSG

Orbital Exenteration Recon Ladder

- **Local options**
  - **Spontaneous granulation**
    - Several months
    - Infection risk
    - Skin contracture
    - Eyebrow ptosis
    - Easy to care for
  - **STSG**
  - **FTSG**

Orbital Exenteration Recon Ladder

- Regional options
  - Temporalis muscle flaps
  - Cervicofacial flaps
  - Temporoparietal fascial
  - Forehead
  - Frontal flaps

Orbital Exenteration Recon Ladder

- Distant options
  - Free tissue transfer flaps
    - Anterolateral thigh
    - Rectus abdominis
    - Latissimus dorsi
    - Radial forearm
    - Lateral arm

Free Tissue Transfer

- Local tissue flaps lack tissue volume

- Prior XRT → local tissue transfer less desirable

Free Tissue Transfer

- Single-stage resection and reconstruction

- Allows ablative surgeon to perform the resection without regard to tissue preservation.

Figure 2. This patient presented with a large neglected basal cell carcinoma of the face. It involved the orbit, maxilla, and a large cutaneous component.
Bibliography