Orbital Considerations in Sinonasal Malignancy

Mindy R. Rabinowitz, MD
Grand Rounds
June 8, 2016
Outline

• Anatomy
• Epidemiology
• Presentation
• Pathology
• Orbital Invasion
• Evaluation
• Management
• Sequela
• Disease-specific management
• Research
Outline

- **Anatomy**
  - Sinus/Orbit
    - Epidemiology
    - Presentation
    - Pathology
    - Orbital Invasion
    - Evaluation
    - Management
    - Sequela
    - Disease-specific management
    - Research
Anatomy

Sinonasal

Superior Turbinate

Middle Turbinate

Inferior Turbinate

Sphenoid

Hard Palate

Nasal Spine
Anatomy

Sinonasal

Lateral Nasal Sidewall
Anatomy

Sinonasal

Lateral Nasal Sidewall

Medial Wall of Maxillary Sinus
Anatomy

Sinonasal

Medial Orbital Wall/
Lamina Papyracea

Ethmoid Air Cells
Anatomy

Sinonasal

Lamina Papyracea
Anatomy

Orbit

Lacrimal Fossa
Anatomy

Orbit

Maxillary
Anatomy

Zygomatic

Orbit
Anatomy

Orbit

Sphenoid
Anatomy

Orbit

Lacrimal
Anatomy

Orbit

Ethmoid
Anatomy

Orbit

Palatine
Anatomy

Orbit

Lamina Papyracea

Lacrimal Fossa
Anatomy

Orbit

- Fronto-ethmoidal suture
- Anterior Ethmoid Artery
- Posterior Ethmoid Artery
- Superior Orbital Fissure
- Inferior Orbital Fissure
- Optic Canal
- 6 mm
- 12 mm
- 24 mm

Fronto-ethmoidal suture
Anatomy

Peri-orbita
Anatomy

Orbit

Periorbita

Lateral canthal tendon
Anatomy

Orbit

Septations

Periorbital Fat
Fat tracks back to orbital apex
Epidemiology

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Epidemiology

Prevalence
- 1% of all malignancies
- ~5% of H&N malignancies

Incidence
- Europeans: 1: 100,000
- Asians: 3: 100,000

SEER Data
- 6730 Non-lymphoreticular malignancies
- 1973-2006

Age
- 6th decade

Gender
- Male (58%)

Race
- White (82%) > Black > “Other”
Rate of Sinonasal Malignancy

Gender

Turner, Reh. 2012
Epidemiology

Race

Turner, Reh. 2012
• Anatomy
• Epidemiology

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Local
- Nasal Obstruction
- Bleeding
- Discharge
- Hyposmia
- Unilateral ETD
- Epistaxis

Regional
- LAD
- Orbital Changes
  - Diplopia
  - Epiphora
  - CN Dysfunction

- Exophthalmos
  - Headache
  - Facial Swelling
  - Numbness

Turner, Reh. 2012
Yang, Hong-gang, Zhen-kun. 2012
Outline

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Sinonasal Malignancies

Epithelial Epidermoid
- Squamous cell ca
- Lymphoepithelial ca
- Carcinomas of lacrimal sac

Mesenchymal
- Solitary Fibrous Tumors
- Liposarcoma
- Fibrosarcoma
- Malignant Fibrous Histiocytoma
- Synovial Sarcoma
- Alveolar Soft Part Sarcoma

Bone
- Osteosarcoma

Lymphoreticular
- Lymphoma
- Extramedullary Plasmacytoma

Cartilage
- Chondrosarcoma

Salavary
- Adenocarcinoma
- Adenoid Cystic Ca
- Mucoepidermoid Ca
- Acinic cell ca
- Carcinoma ex-pleomorphic adenoma

Vasoform
- Angiosarcoma

Muscle
- Rhabdomyosarcoma
- Leiomyosarcoma

Neuroectodermal
- Malignant Peripheral Nerve Sheath Tumor
- Meningioma
- Neuroendocrine Carcinoma
- SNUC
- Ewings
- Olfactory Neuroblastoma
- Mucosal Melanoma

Metastases
<table>
<thead>
<tr>
<th>Sinonasal Malignancies</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Epithelial Epidermoid</strong></td>
</tr>
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Salavary

Respiratory Epithelial
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Metastases
- Squamous Cell Carcinoma
Sinonasal Malignancies

80% Sinonasal Malignancies

60-70 years of age

M > F

Risk Factors:

- Nickle
- Chromium
- Bantu snuff
- Hydrocarbons
- Radium
- XRT
- IP

5 year survival: 60%

Surgery, Chemo, XRT

Loevelner, Sonners 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
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Metastases
- Adenocarcinoma
Sinonasal Malignancies

Adenocarcinoma

Ethmoid Sinuses

Leather & Wood Workers

Advanced at Presentation:
50% cribiform erosion

Tx: Surgery → XRT

Loevner, Sonners 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
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Metastases
- Adenoid Cystic Carcinoma
Sinonasal Malignancies

Adenoid Cystic Carcinoma

- Most common salivary neoplasm
- Location:
  - Maxillary > N.C. > Sphenoid/Frontal
- Peri-neural spread
  - Extension → orbit & intracranial
  - Difficult to clear
- Treatment:
  - Surgical rsxn
  - Adjuvant XRT
- 50% Distant Mets → Lung, Brain, Bones

Loechner, Sonners 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
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Metastases
- Rhabdomyosarcoma
Sinonasal Malignancies

Rhabdomyosarcoma

- Embryonal, Alveolar, Pleomorphic
  - Adults: 2-5%
  - Children: 60%

- 35% Head and Neck
- 20%
- Nasal Cavity, Nasopharynx, Sinuses
  - Adults: Ethmoids

Treatment: Surgery, Chemo/XRT

Loevinger, Sonners 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
Sinonasal Malignancies

Epithelial Epidermoid
- Squamous cell carcinoma

Salivary

Sinonasal Undifferentiated Carcinoma (SNUC)

Cartilage
- Lymphoma
- Extramedullary Plasmacytoma
- Chondrosarcoma

Metastases
- Neuroendocrine Carcinoma
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Sinonasal Malignancies

Sinonasal Undifferentiated Carcinoma

Derived from Schneiderian epithelium

Mean age of dx: 6th decade

Aggressive neoplasms
  • Advanced presentation
    • 20-30% Cervical LAD
  • Metastatic disease

Rapid growth
  • Site of origin?

Treatment Controversial:
  • Chemo/XRT w/ surgical salvage
  • Surgery → Chemo/XRT

Loevoer, Sonders 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
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Olfactory Neuroblastoma

Metastases
Sinonasal Malignancies

Olfactory Neuroblastoma

- Arise from olfactory neuroepithelium
- Bimodal
  - 2\textsuperscript{nd}-3\textsuperscript{rd} decade
  - 6-7\textsuperscript{th} decade
- Cross cribiform \rightarrow Intracranially
- Treatment:
  - Craniofacial rsxn + XRT
- Long natural history
  - Locoregional recurrence
    - Up to 10 years
- Long term follow up necessary

Loevner, Sonners 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
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Metastases
- Mucosal Melanoma
Sinonasal Malignancies

~20% Head & Neck

~2-5% Sinonasal Cavity

Nasal Septum

Turbinate → 80% Maxillary Sinus

Male > Female

5-7th Decade

Spread by local invasion

LN Metastasis → 6% of cases

Treatment:

- Surgery ± XRT
- Local Failure: 50%
- Mean Survival: ~2 years

Loevner, Sonners 2002
Raghavan, Phillips, 2007
Lung, Howard, Wei. 2014
• Anatomy
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• Orbital Invasion

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• Disease-specific management
• Research

• Epidemiology
• Presentation
• Mechanism of Action
• Definition
• Prognosis
Orbital invasion by malignancies

- Varies with:
  - Site of origin
  - Histology
  - Tumor Aggressiveness

- Diagnosed at advanced stage

- Orbital Involvement:
  - 60-80%: Maxillary & Ethmoid
  - 30-50% Periorbital Involvement

Bleier, Lin. 2012
Suarez, Ferlito, Dpath et. al. 2007
Ocular Symptoms

- 62% Ethmoidal Tumors
- 46% Nasal Cavity Tumors
- Motility Disturbance (31%)
- Afferent System Dysfunction (29%)
- Proptosis (29%)
- Eyelid Abnormalities (19%)
- Dystopia (17%)
- Lacrimal Outflow Obstruction (5%)
- Numb/Pain

Bleier, Lin. 2012
Loevner, Sonners. 2002
Essig, Newman, Levine. 2007
Suarez, Ferlito, Dpath et al. 2007
Singh, Eskander, Huang, et al. 2013
All 4 sinuses surround the orbit...
Mechanism of Action

All 4 sinuses surround the orbit...

Bleier, Lin. 2012
McCary, Levine. 1995
Singh, Eskander, Huang, et.al. 2013
All 4 sinuses surround the orbit...
Opportunities for orbital extension

- Multiple foramina
- Perforating nerves/ vessels
- Thin bones
  - Lacrimal fossa
  - Lamina papyracea
- Periorbita

No natural intra/extra-conal barriers
“Orbital Invasion”

Orbital wall erosion?

Periorbital Involvement?

Orbital Soft Tissues?

Orbital Invasion

Curtin, Rabinov. 1998
Kim, Lee, Lee, et.al. 2015
Carrau, Segas, Nuss, et.al. 1999
Suarez, Ferlito, Dpath et. al. 2007
Iannetti, Valentini, Rinna, et.al. 2005
Singh, Eskander, Huang, et.al. 2013
Periorbita

- “True protector” of orbit
- Intact despite bone erosion
- Defines orbital invasion?
- Degree of invasion
  - Partial vs. Full thickness?
Orbital Invasion - 35-75% of cases

Discordant definitions

McCary et al

- A: Adj to orbit, no wall
- B: Wall erosion, no globe displacement
- C: Wall erosion & infiltration, no periorbita
- D: Invades orbit & periorbita

Ianetti et al

- I: Wall erosion
- II: Orbital fat
- III: MR, globe, ON, palpebral skin

No Agreement

References:
Curtin, Rabinov. 1998
Kim, Lee, Lee, et.al. 2015
Carrau, Segas, Nuss, et.al. 1999

Suarez, Ferlito, Dpath et. al. 2007
Ianetti, Valentini, Rinna, et.al. 2005
Singh, Eskander, Huang, et.al. 2013
Prognosis

Poor Prognosis

Ganly et al 2005

334 pts CFR for ethmoid tumors

LACK of orbital invasion...

- Predict recurrence free, disease specific and overall survival
  - DSS:
    - + Orbit: 41%
    - - Orbit: 75%

Orbital soft tissue involvement

- Independent factor influencing survival

Invasion limited to periorbita

- Survival unchanged

Bleier, Lin. 2012
Ganly, Patel, Singh et al. 2005
Suarez, Ferlito, Dpath et. al. 2007
Outline

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Evaluation

- Management
- Sequela
- Disease-specific management
- Research

- Imaging
- Definitive

Disease-specific management
• No symptoms ➞ No invasion
• CT & MRI important!

Penetrate bony orbit; possibly periorbita
Erode bone only
Directly abut the orbit w/o bone invasion
Orbital soft tissues

Useful for distinguishing...
Imaging

Eisen et al. 2000
Predict Periorbital Invasion

**Criteria**

- **Relationship to Periorbita**
  - Abutting
  - Displacing
  - Bowing

- **Interface: Tumor & Periorbita**
  - “nodular”
  - “smooth”

- **Orbital Fat Invasion**
  - Stranding in extraconal fat

**Extraocular Muscles**
- Displaced
- Enlarged
- Abnl intensity

**Orbital Bone Integrity**

**Nasolacrimal Invasion**
- Tumor in duct or sac

**Conclusions**
- No one criterion had accuracy >79%
- ≥ 6 criteria predicted orbital invasion w/ accuracy 72%.
- Adjacent to periorbita: most sensitive (90%), low specificity (29-44%)
- EOM enlargement: most specific (94%)
- Orbital Fat Involvement: specific, high PPV, but less sensitive
CT more accurate than MRI

Strength of CT

• Evaluate bone, fat interface

Difficulty distinguishing tumor...

• Compresses vs. invades periorbita

Loevner, Sonners. 2002
Kim, Lee, Lee, et al. 2006
Erosion of lateral nasal wall and frontal sinus floor

Thinning & bowing
Medial orbital wall

Loevner, Sonners. 2002
Kim, Lee, Lee, et.al. 2006
Invasion of the periorbita:

- Results in loss of signal
Loss of fat signal

Abnormal signal in fat

Normal bright fat on T1

Tumor Nodularity

Mass Effect of Medial Rectus

Loechner, Sonners, 2002
Kim, Lee, Lee, et al. 2006
Eisen, Yousem, Loechner, et al. 2000
Singh, Eskander, Huang et al. 2013
Imaging

Tumor Nodularity

Loevner, Sonners. 2002
Kim, Lee, Lee, et.al. 2006
Intra-operative frozen section
Outline

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- Evaluation

Management

- Sequela
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- Research

- Controversy
- Surgery
- Radiation/Chemo
Management

- Surgery
- XRT
- Chemo
Management

Surgery
- Exenteration
- Debulk

XRT

Chemo
Points of Contention

1. Oncological Safety of Orbital Preservation

2. Functional Outcome in Preserved Eyes.
1. Involvement of the orbital apex
2. Non-resectable full-thickness invasion through periorbita into retrobulbar fat
3. Extension into the EOM
4. Invasion of the bulbar conjunctiva
5. Lid involvement beyond reasonable hope for reconstruction

• Study over 200 pts
• Local control rate:
  • 79% with exenteration
  • 14% without exenteration

Probably unwise to conclude that the orbit can be spared in all patients
Orbital Exenteration

Support for

Anderson (1996)

- 32% incidence of non-functional eye
- 58 patients- anterior CFR, orbit preservation, adjuvant XRT

“Strong consideration for orbital exent based on anticipated poor functional outcomes”

Rajapurkar (2013)

- 19 total/radical maxillectomy
  - 4/19- orbital floor + periorbita + Fat (orbit preserved)
  - ¾- orbital + margins (despite XRT)
  - Recurred

If cant ensure complete negative orbital margins, exent may be safer oncologically
Orbital Exenteration

Contraindications
- Brain?
- Cavernous Sinus
- Carotid Artery
- Bilateral Optic n.
- Optic Chiasm

Support
Against

Psychosocial
- 27 patients → Orbital Preservation
  - All were glad eye was spared

The cosmetic, functional, and psychologic consequences of loss of an eye mandate that every effort should be made to preserve the eye as long as oncologic safety is not compromised.”
### Orbital Exenteration

#### Support Against

Reyes, Mason, Solares, Bush, Carrau. 2015

<table>
<thead>
<tr>
<th>Source</th>
<th>Indication for evisceration</th>
<th>5-year survival evisceration, %</th>
<th>5-year survival preservation, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wu et al^{17}</td>
<td>Invasion medial orbital wall^a</td>
<td>27.3</td>
<td>34.8</td>
</tr>
<tr>
<td>Lund et al^{19}</td>
<td>Transgression of the periosteum^b</td>
<td>29</td>
<td>26</td>
</tr>
<tr>
<td>Imola et al^{20}</td>
<td>Orbital fat involvement, extraocular muscle invasion, orbital apex, or eyelid invasion^c</td>
<td>46</td>
<td>53</td>
</tr>
<tr>
<td>Iannetti et al^{18}</td>
<td>Invasion of the medial rectus muscle, optic nerve, ocular bulb, or the skin overlying the eyelid^c</td>
<td>62.2</td>
<td>63.5</td>
</tr>
</tbody>
</table>

No strong evidence favoring preservation vs. exenteration

**Criticisms:**

- Tumor histology not taken into account.
- Effect may be limited to SCC, Adenoca
- Different indications for exenteration
- Preserved: not description of what resected
Orbital Exenteration

- “Starved” the day of the operation
- Held down by 2 assistants
- No anesthesia
- Pass needle & silk thread through globe
  - Exerting forward traction
- Cut attachments to eye with curved knife
- Bandage placed
  - Enriched with sulfered spelter and brandy

Bartisch

1583

Straub. 1988
McCary, Levine. 1995
Imola, Schramm. 2002
Perry, Levine, Williamson, et al. 1988
Orbital Exenteration

Involves removal of the soft tissue contents of the orbit, including the globe.

**Total:**
- globe, eyelid, conjunctiva, orbital contents, periorbita

**Extended:**
- bony orbital walls, paranasal sinuses, ± intracranial tissue

**Subtotal:**
- globe, conjunctiva, EOM
Orbital Exenteration

Orbicularis

Periosteum
Orbital Exenteration

Lacrimal sac (cut)
Orbital Exenteration

Inferior orbital fissure
Management

**Surgery**
- Exenteration
- Debulking

**XRT**

**Chemo**
Eye preservation could be attempted in any malignancy that invaded the orbital soft tissues with penetration through the periorbita provided that it could be easily and completely dissected away from the orbital fat.

TABLE IV.
Patterns of Local Recurrence According to Primary Tumor Site in Patients Treated With Orbital Preservation versus Orbital Exenteration.

<table>
<thead>
<tr>
<th>Histologic Subtype</th>
<th>No.</th>
<th>-Orbital Disease</th>
<th>+Orbital Disease</th>
<th>Total</th>
<th>No.</th>
<th>Local Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCCa</td>
<td>18</td>
<td>4</td>
<td>2</td>
<td>6 (11.1)*</td>
<td>6</td>
<td>2 (16.7)</td>
</tr>
<tr>
<td>SNUC</td>
<td>6</td>
<td>1</td>
<td>1</td>
<td>2 (3.7)</td>
<td>2</td>
<td>1 (8.3)</td>
</tr>
<tr>
<td>Adenomatous</td>
<td>10</td>
<td>2</td>
<td>1</td>
<td>3 (5.6)</td>
<td>3</td>
<td>1 (8.3)</td>
</tr>
<tr>
<td>Sarcoma</td>
<td>11</td>
<td>3</td>
<td>—</td>
<td>3 (5.6)</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>Other</td>
<td>9</td>
<td>2</td>
<td>—</td>
<td>2 (3.7)</td>
<td>—</td>
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</tr>
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<td>TOTAL</td>
<td>54</td>
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</tbody>
</table>

- Orbital disease = local recurrence in the sinonasal cavity remote from the original site of orbital involvement.
+ Orbital disease = local recurrence in the sinonasal cavity with involvement of the original orbital site.
*Values in parentheses are percentages.

No statistically significant difference in recurrence rates.
Preservation of the orbit, when the full thickness of periorbita is not invaded, does not downgrade outcome and therefore supports a trend toward orbital preservation.

Disease-free survival in patients with orbital invasion

<table>
<thead>
<tr>
<th>Study</th>
<th>Orbital Preservation</th>
<th>Exenteration</th>
<th>Significance (P)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Som et al. (1974)</td>
<td>1/2 (50%)</td>
<td>3/27 (11%)</td>
<td>&gt;.05</td>
</tr>
<tr>
<td>Perry et al. (1988)</td>
<td>8/23 (35%)</td>
<td>24/88 (27%)</td>
<td></td>
</tr>
<tr>
<td>Xuexi et al. (1995)</td>
<td>5/9 (56%)</td>
<td>6/12 (50%)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>14/34 (41%)</td>
<td>35/131 (37%)</td>
<td></td>
</tr>
</tbody>
</table>

Local recurrence in patients with orbital invasion

<table>
<thead>
<tr>
<th>Study</th>
<th>Orbital Preservation</th>
<th>Exenteration</th>
<th>Significance (P)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Som et al. (1974)</td>
<td>-</td>
<td>22/27 (81%)</td>
<td></td>
</tr>
<tr>
<td>Perry et al. (1988)</td>
<td>1/2 (50%)</td>
<td>1/4 (25%)</td>
<td></td>
</tr>
<tr>
<td>Xuexi et al. (1995)</td>
<td>2/23 (9%)</td>
<td>11/88 (13%)</td>
<td></td>
</tr>
<tr>
<td>McCary et al. (1996)</td>
<td>1/5 (20%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Carrau et al. (current study)</td>
<td></td>
<td>8/16 (50%)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Survival: Sinonasal Malignancy

5-Year Survival: Sinonasal Malignancy

Not specific to orbital invasion

Turner, Reh. 2012
University of Virginia Protocol (SNUC, Esthesio)

- Preop XRT
  - Effort to spare globe, lacrimal gland
- Chemo?
- Salvage surgery

- **33 patients: Orbital invasion**
  - Invaded bone & displaced periorbita
  - Grossly invaded the orbit & periorbita
- No eyes removed
- 5/33: Local recurrence
  - 1/5: in orbit

Pre-op XRT

- Reduce tumor size
- Tumor in orbit replaced by scar
  - Easily dissected from fat

McCary, Levine. 1995
McCary, Levine, Cantrell. 1996
Xuei, Pingzhang, Yongfa. 1995
Algorithm for Managing Orbit

**Biopsy/Imaging**

- **Lamina Eroded**
  - **Through Periosteum**
    - **Orbital Clearance**
  - **Adjacent to periosteum**
    - **Resect Periosteum Frozens**
- **Lamina Intact**
  - **Periosteum clear**
    - **Orbital Preservation**

Suarez, Ferlito, Lund, et.al. 2008
Outline

- Anatomy
- Epidemiology
- Presentation
- Pathology
- Orbital Invasion
- Evaluation
- Management

- Sequela
  - Disease-specific management
  - Research
Points of Contention

Oncological Safety of Orbital Preservation

Functional Outcome in Preserved Eyes
Sequela of Function

Surgery alone

radiation alone

Surgery & radiation
Surgery alone

Imola, Schramm 2002

- Functional without impairment: 54%
- Functional with impairment: 37%
- Non-Functional: 9%

Most Common:
- Lack of adequate reconstruction
- Diplopia

“Functional” day-to-day use of the eye (VA, globe position, lacrimal integrity, etc..)
Loss of Ocular Function

- Ipsilateral Orbit: 79%
- Contralateral Orbit: 5%

Stern, Goepfert, Clayman, et.al. 1993
Loss of Ocular Function

Overall Incidence of Functional Ocular Sequelae and the Influence of Radiation Therapy in the Group of Patients Treated With Orbital Preservation.

<table>
<thead>
<tr>
<th>Ocular Sequelae</th>
<th>Occurrence in Orbital Preservation Group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Overall (54)</td>
</tr>
<tr>
<td></td>
<td>Radiation Therapy (39)</td>
</tr>
<tr>
<td></td>
<td>No Radiation Therapy (15)</td>
</tr>
<tr>
<td>Ectropion</td>
<td>11 (20)*</td>
</tr>
<tr>
<td></td>
<td>9 (23)</td>
</tr>
<tr>
<td></td>
<td>2 (13)</td>
</tr>
<tr>
<td>Blepharitis/conjunctivitis</td>
<td>18 (33)</td>
</tr>
<tr>
<td></td>
<td>15 (38)</td>
</tr>
<tr>
<td></td>
<td>3 (20)</td>
</tr>
<tr>
<td>Corneal exposure keratopathy</td>
<td>6 (11)</td>
</tr>
<tr>
<td></td>
<td>5 (13)</td>
</tr>
<tr>
<td></td>
<td>1 (7)</td>
</tr>
<tr>
<td>Epiphora</td>
<td>7 (13)</td>
</tr>
<tr>
<td></td>
<td>2 (5)</td>
</tr>
<tr>
<td></td>
<td>5 (33)</td>
</tr>
<tr>
<td>Dryness</td>
<td>5 (9)</td>
</tr>
<tr>
<td></td>
<td>5 (12.8)</td>
</tr>
<tr>
<td>Optic atrophy</td>
<td>2 (4)</td>
</tr>
<tr>
<td></td>
<td>2 (5)</td>
</tr>
<tr>
<td>Cataract formation</td>
<td>4 (8)</td>
</tr>
<tr>
<td></td>
<td>4 (10)</td>
</tr>
</tbody>
</table>

*Values in parentheses are percentages.
Sequela of Function

Surgery & radiation

McCary 1996

- 29/33-
  - Pre-op XRT
  - Rxn of periorbita

- 55%: No ophthalmic problems
- 45%:
  - Exposure Keratitis (6 pts)
  - Motility Disturbance (6 pts)
    - 5 pts: transient or asymptomatic
  - Cataracts
  - Ectropion
  - Dysotopia
  - Enophthalmos

Bleier 2012, Suarez 2008

- Increased risk of:
  - Optic atrophy
  - Cataracts
  - Excessive dryness
  - Ectropion
Sparing the soft tissues of the orbit when the periorbita have not been deeply transgressed by tumor generally does not appear to adversely affect local control.
Outcomes...

Confounding factors...

Selection Bias

- More advanced tumors → Exenteration
- More favorable tumors → Preservation

Tumor Histology

- Plays significant role in outcome
<table>
<thead>
<tr>
<th>Histologic Subtype</th>
<th>No.</th>
<th>−Orbital Disease</th>
<th>−Orbital Disease</th>
<th>Total</th>
<th>No.</th>
<th>−Orbital Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCCa</td>
<td>18</td>
<td>4</td>
<td>2</td>
<td>6 (11.1)*</td>
<td>6</td>
<td>2 (16.7)</td>
</tr>
<tr>
<td>SNUC</td>
<td>6</td>
<td>1</td>
<td>1</td>
<td>2 (3.7)</td>
<td>2</td>
<td>1 (8.3)</td>
</tr>
<tr>
<td>Adenomatous</td>
<td>10</td>
<td>2</td>
<td>1</td>
<td>3 (5.6)</td>
<td>3</td>
<td>1 (8.3)</td>
</tr>
<tr>
<td>Sarcoma</td>
<td>11</td>
<td>3</td>
<td>−</td>
<td>3 (5.6)</td>
<td>3</td>
<td>−</td>
</tr>
<tr>
<td>Other</td>
<td>9</td>
<td>2</td>
<td>−</td>
<td>2 (3.7)</td>
<td>2</td>
<td>−</td>
</tr>
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*Values in parentheses are percentages.

Low numbers of each histology
Conclusions...

Periorbita not deeply transgressed

✧ Does not affect local control/survival

✧ Preserve orbit

Outcomes less clear when fat is invaded

Surgery/XRT- Preserved eye with variable level of function

Tumor histology/behavior matters

Imola, Schramm, 2002
Outline

• Anatomy
• Epidemiology
• Presentation
• Pathology
• Orbital Invasion
• Evaluation
• Management
• Sequela
• Disease-specific management

• Research
Jefferson Data

Goals

1. Evaluate recurrence and survival rates based on tumor histology for orbital preservation vs. exenteration

2. Stratify based on extent of tumor resection & degree of invasion
   • Attention to orbital fat
<table>
<thead>
<tr>
<th>Tissue Type</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inv pap w/ SCC</td>
<td>6</td>
</tr>
<tr>
<td>SCC</td>
<td>15</td>
</tr>
<tr>
<td>BCC</td>
<td>2</td>
</tr>
<tr>
<td>Melanoma</td>
<td>3</td>
</tr>
<tr>
<td>Esthesio</td>
<td>5</td>
</tr>
<tr>
<td>Poorly diff/undiff</td>
<td>13</td>
</tr>
<tr>
<td>Spindle Cell</td>
<td>2</td>
</tr>
<tr>
<td>SNUC</td>
<td>2</td>
</tr>
<tr>
<td>ACC</td>
<td>2</td>
</tr>
</tbody>
</table>

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<tr>
<th>Tissue Type</th>
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</tr>
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<tbody>
<tr>
<td>MucoEp</td>
<td>1</td>
</tr>
<tr>
<td>Hemangioperi</td>
<td>1</td>
</tr>
<tr>
<td>AdenoCA</td>
<td>5</td>
</tr>
<tr>
<td>Sarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Alveolar Rhabdo</td>
<td>2</td>
</tr>
<tr>
<td>Sebaceous CA</td>
<td>1</td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>Myoepithelioma</td>
<td>1</td>
</tr>
<tr>
<td>Plasmacytoma</td>
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</tbody>
</table>
Acknowledgements

Otolaryngology
Marc Rosen
Gurston Nyquist

Oculoplastics
Michael Rabinowitz

Medical Students
Kriner Gill
Varun Patel
Krystal Park
• References


Questions?