COURSE 8
Lumps and Bumps: Ocular Oncology

COPE Course 42486-SD

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Eyelid and Orbital “Lumps and Bumps”

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Benign Eyelid Lesions

• Nodules
  • Chalazion
  • Acute hordeolum
  • Molluscum contagiosum
  • Xanthelasma

• Tumors
  • Actinic Keratosis
  • Seborrheic Keratosis
  • Papilloma
  • Pyogenic granuloma
  • Melanocytic Nevus
  • Neurofibroma
  • Keratoacanthoma

• Cysts
  • Hidrocystoma
  • Sebaceous cyst
  • Cyst of Zeiss

Nodules

What Is It?
• Cyst within tarsal plate caused by inflammation of blocked meibomian gland

What Does It Look Like?
• Raised, round, painless, firm lesion typically on the upper eyelid

Who Gets It?
• M=F, all ages
• Predisposing conditions: blepharitis, rosacea

Medical Management
• Warm compresses
• Topical antibiotic/steroid ointment/drops
• Steroid injection

Surgical Management
• Incision and drainage
• Excision

Chalazion

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• Cyst within tarsal plate caused by inflammation of blocked meibomian gland

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• Who Gets It?
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External Hordeolum

- **What is it?**
  - Staphylococcus abscess of lash follicle and associated gland of Moll or Zeiss
- **What Does it Look Like?**
  - Tender bump at lid margin with associated swelling
- **Who Gets it?**
  - M=F, all ages
  - Predisposing conditions: poor nutrition, poor hygiene, history of recurrent infections

Molluscum Contagiosum

- **What is it?**
  - Viral induced lesion on the skin or mucous membranes
  - May cause chronic follicular conjunctivitis or superficial keratitis
- **What Does it Look Like?**
  - Painless, waxy umbilicated nodule
- **Who Gets it?**
  - Children
  - Patients with AIDS - multiple

Medical Management

- DO NOT forcefully rupture – may cause bacterial spread
- Topically or oral antibiotics

Surgical Management

- Typically not necessary
- Excision after full course antibiotics

Xanthelasma

- **What is it?**
  - Deposition of lipid or cholesterol beneath skin usually on or around eyelids
- **What Does it Look Like?**
  - Sharply demarcated yellowish plaques; bilateral; medial

Medical Management

- OTC salicylic acid
- Tretinoin cream
- Requires several months of treatment and may cause discomfort

Surgical Management

- Cryosurgery
- Curettage
Who Gets It?
- Elderly
- Mediterranean or Asian descent
- Hypercholesterolemia

Medical Management
- Deep TCA chemical peels
- Lasers

Surgical Management
- Cryosurgery
- Excision

Cysts
- Hidrocystoma (Moll’s gland cyst)
  - Eyelid margin
  - Translucent
- Sebaceous Cyst
  - Often at medial canthus
  - “Cheesy” contents
- Cyst of Zeiss
  - Anterior lid margin
  - Opaque/white

Benign Tumors

Actinic Keratosis
- What Is It?
  - Slow growing, pre-malignant keratinization of skin
  - Results from excessive sun exposure
- What Does It Look Like?
  - Flat, scaly, rough plaque;
  - May be multiple
  - Rare on eyelid

Seborrheic Keratosis
- What Is It?
  - Slow growing, pre-malignant keratinization of skin
  - Results from excessive sun exposure
- What Does It Look Like?
  - Flat, greasy, often pigmented
  - “Stuck-on” appearance
Who Gets It?

- Elderly

Medical Management

- None

Surgical Management

- Biopsy for definitive diagnosis

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**Papilloma**

- What Is It?
  - Proliferation of fibrovascular tissue covered by irregular keratinized squamous epithelium
  - May be caused by viral infection (HPV)

- What Does It Look Like?
  - "Skin tag"
  - Pedunculated or broad base

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Who Gets It?

- Anyone

Medical Management

- None

Surgical Management

- Biopsy for definitive diagnosis

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**Pyogenic Granuloma**

- What Is It?
  - Overgrowth of vascular tissue

- What Does It Look Like?
  - Fast-growing, pinkish lesion
  - Pedunculated or flat based

- Who Gets It?
  - History of recent surgery or trauma

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Medical Management

- Topical or intralesional steroid

Surgical Management

- Excision
  - Beware of recurrence

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**Melanocytic Nevus**

- What Is It?
  - Tumor composed of melanocytes; congenital or acquired

- What Does It Look Like?
  - Junctional: Uniform brown macule or plaque
  - Compound: Uniform, light to dark brown, raised papule
  - Intradermal: Papillomatous with little to no pigment. Associated with dilated vessels and protruding lashes

- Who Gets It?
  - May become more pigmented in puberty
Intradermal Junctional Compound

- Elevated
- May be amelanotic
- No malignant potential

- Elevated
- Usually pigmented
- Low malignant potential

Has components of both intradermal and junctional

Surgical Management

- Excisional biopsy

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Neurofibroma

- What Is It?
  - Abnormal proliferation of Schwann cells and fibroblasts
  - Plexiform type found on the eyelid

- What Does It Look Like?
  - Large, “s-shaped” elevated lesion on upper eyelid
  - “Bag of worms” texture

- Who Gets It?
  - Solitary lesion in adults
  - 25% associated with Neurofibromatosis – 1 (multiple)

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Medical Management

- None

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Surgical Management

- Excision

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Keratoacanthoma

- What Is It?
  - Uncommon pre-malignant proliferation of squamous epithelium

- What Does It Look Like?
  - Fast growing, keratin-filled crater with rolled edges
  - May spontaneously involute within 1 year

- Who Gets It?
  - Elderly, fair-skinned
  - Immunosuppressed

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Medical Management

- None

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Surgical Management

- Occasionally cryosurgery
  - Excision

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Malignant Eyelid Lesions

- Basal cell carcinoma
- Squamous cell carcinoma
- Sebaceous cell carcinoma
- Melanoma
- Merkel cell carcinoma
- Kaposi sarcoma
Basal Cell Carcinoma

- **What Is It?**
  - Most common eyelid cancer
  - Slow growing, locally destructive proliferation of basal cells
- **What Does It Look Like?**
  - Usually on lower eyelid; destruction of normal architecture
  - Nodular type: pearl like, with dilated blood vessels on surface
  - Ulcerative type: central ulcer with raised pearly edges
  - Sclerosing type: lateral, hardened, infiltration beneath the epidermis. May be confused with chronic blepharitis

Who Gets It?
- Elderly, fair-skinned
- Excessive sun exposure

Treatment
- Biopsy
- Excision with 3-4mm clear margins

Squamous Cell Carcinoma

- **What Is It?**
  - Aggressive cancer, can arise de novo or from AK or keratoacanthoma
  - Can metastasize to local lymph node in 20%
- **What Does It Look Like?**
  - Scaly with irregular boarders; may bleed
  - Nodular type: keratinized nodule; develops erosions and fissures
  - Ulcerating type: everted boarders with red, well defined base
  - Cutaneous horn: invasive growth underlies keratin horn

Who Gets It?
- Elderly
- Fair skinned, un exposure, immunosuppressed

Treatment
- Excision with 3-4mm clear margins
- +/- cryotherapy, radiation
- Can be fatal if untreated

Sebaceous Cell Carcinoma

- **What Is It?**
  - Slow growing cancer arising from meibomian glands, glands of Zeiss or caruncle
  - Usually on upper eyelid
- **What Does It Look Like?**
  - Can appear similar to chalazion or chronic blepharitis
  - May have yellowish material internally
  - Nodular type: hard, painless, immobile nodule; chalazion-like
  - Spreading type: thickened lid margin, loss of lashes; blepharitis-like

Lower Eyelid Reconstruction After Cancer Removal
Who Gets It?
- Females, elderly (60's – 70's)

Treatment
- Cryotherapy and surgical excision are the standard treatments
- Recurrence is as high as 33%
- Mortality rate is 5-10%

Melanoma
- What Is It?
  - Highly fatal cancer arising from melanocytes
- What Does It Look Like?
  - Variable colors (blue, black);
  - 50% are amelanotic
  - Asymmetric, irregular with indistinct boarders
  - Destruction of local anatomy and loss of lashes

Who Gets It?
- Caucasians, sun exposure
- More advanced in dark-skinned people
- Increasing incidence in people in their 20's

Treatment
- Wide surgical excision with up to a 1 cm margin
- Local lymph node dissection if more than 1.5 mm deep
- Close follow-up

Merkel Cell Carcinoma
- What Is It?
  - Rare, highly aggressive, rapidly growing neuroendocrine tumor
- What Does It Look Like?
  - Red, purple or violet well-defined nodule
  - Overlying skin is intact

Who Gets It?
- Caucasians, elderly (average age 75)
- Sun exposure, immunocompromised

Treatment
- CT and/or MRI imaging used to evaluate systemic spread (many have mets at time of diagnosis)
- Excision with wide margins (3cm if possible)
- Chemotherapy and/or radiotherapy depending on spread
- 2 year mortality rate of 30-50%

Kaposi’s Sarcoma
- What Is It?
  - Vascular tumor caused by HHV-8
- What Does It Look Like?
  - Erythematous or violaceous patch, path or nodule
Who Gets It?
- Middle aged men
- Mediterranean or African descent
- AIDS

Treatment
- Excision, cryotherapy, intralesional injections of vinblastine, radiotherapy, topical immunotherapy (Imiquod)
- Extensive disease requires chemo and immunosuppression

Orbital Tumors

Benign Orbital Tumors
- Bone adenoma
  - Osteoma
  - Fibrous dysplasia
- Well-delineated
  - Cavernous Hemangioma
  - Hemangiopericytoma
  - Dermoid cyst
  - Mucocele
  - ON tumors
- Diffuse
  - Lymphangioma
  - Benign reactive lymphoid hyperplasia
  - Lacrimal gland (pleomorphic)

Osteoma
- What Is It?
  - Benign skeletal neoplasm of unknown etiology
  - In the orbit, typically involves the frontal and ethmoid bones
  - May cause pain, proptosis, decreased vision, or diplopia

What Does It Look Like?
- Radiographically, these tumors are well-circumscribed with dense cortical sclerosis surrounding a radiolucent nidus.
- Grossly, the lesion has a glistening, white to pink color and is either smooth or with rounded protuberances

Who Gets It?
- Younger patients, typically found incidentally

Treatment
- Excisional biopsy

Cavernous Hemangioma
- What Is It?
  - Benign, noninfiltrative, slowly progressive vascular neoplasm
  - Composed of endothelial-lined spaces surrounded by a well-delineated fibrous capsule
  - Most common benign orbital tumor in adults
  - Typically located intraconally
What Does It Look Like?
- Presents as slowly progressive, painless proptosis
- May have EOM disturbance, induced hyperopia, elevated IOP, choroidal folds, or decreased vision
- On CT scan: well-circumscribed, homogenous mass slightly hyperdense to muscle, located intraconally.

Who Gets It?
- Middle-aged adults, F>M

Treatment
- May be observed as long as not comprising the eye
- Orbital excision indicated for growth, optic ON compression, exposure keratopathy, or evidence of vision loss.

Dermoid Cyst
- What Is It?
  - Congenital tumor consisting of keratinized epithelium and adnexal structures (hair follicles, sweat glands, and sebaceous glands)
- What Does It Look Like?
  - Egg-shaped, smooth, firm mass under the skin adjacent to bone
  - CT scan: well-circumscribed lesion with a hyperdense wall and hypodense contents. Bony remodeling is present in 85% of cases.

Who Gets It?
- Typically found in children
- Adults present with deeper, more posteriorly located tumors

Treatment
- If small, may be observed
- Complete excision – beware of inflammatory response if ruptures or recurrence/abscess if incompletely removed

Mucocele
- What Is It?
  - Mucous or fluid-filled cyst arising from the ethmoid or frontal sinuses that subsequently invades the orbit
- What Does It Look Like?
  - Slowly progressive displacement of eye (may be axial or non-axial proptosis)
Who Gets It?
Middle-age
History of chronic sinus disease or facial trauma

Treatment
MRI
Drainage procedure—send fluid for culture and cytology
Surgery—orbitotomy and sinusctomy; removal of as much of the cyst and its lining as possible

Lymphangioma

What Is It?
Diffusely infiltrating benign nonencapsulated vascular tumor

What Does It Look Like?
Fullness in the superior or nasal quadrant with acute proptosis after minor head trauma or respiratory URI
>50% affect anterior structures causing bluish discoloration of or blood vessels within the eyelid skin.
May bleed into itself causing cysts of blood, called chocolate-cysts

Who Gets It?
Any age

Treatment
If small, may be observed
Complete excision—beware of inflammatory response if ruptures or recurrence/abscess if incompletely removed

Pleomorphic Adenoma

What Is It?
Neoplastic proliferation of epithelial cells in the lacrimal gland

What Does It Look Like?
Unilateral slowly progressive proptosis (inferomedial displacement)

Who Gets It?
Middle age

Treatment
CT—solid (can be heterogeneous), well defined, round or oval, occasional calcification, and bony remodeling
Complete surgical excision (incomplete removal can result in recurrence or malignant transformation)

Malignant Orbital Tumors

Bone
• Osteosarcoma
• Metastasis

Well-delineated
• Metastasis
• Melanoma

Diffuse
• Lymphoma
• Lacrimal gland
• Pleomorphic adenocarcinoma
• Adenoid cystic carcinoma
Osteosarcoma

- What is it?
  - Very rare orbital tumor that forms osteoid
  - Arises from soft tissue (NOT bone)

- What Does It Look Like?
  - Rapidly growing, painless mass and/or proptosis
  - CT – well-circumscribed calcified mass

Who Gets It?

- Patients > 60 years old
- History of immunosuppressive or prior radiation

Treatment

- Undefined (radical surgery, radiation, aggressive chemotherapy)
- Poor prognosis, unless well-differentiated histologically

Metastasis

- What is it?
  - Metastasis from breast carcinoma > melanoma > prostatic cancer

- What Does It Look Like?
  - Proptosis, strabismus and vision loss are the common clinical signs
  - CT- solid enhancing mass located within the orbital fat or enlargement of an extraocular muscle

Who Gets It?

- History of cancer; however, the orbit is the first presentation in 15%

Treatment

- Diagnosis confirmed with fine needle aspiration biopsies, serological studies, and molecular biology techniques
- Multi-disciplinary and multiple modalities - radiotherapy, chemotherapy, hormone therapy, surgery, and immunotherapy
- Survival after diagnosis is 1.5 years on average, independent of the histological type

Primary Melanoma

- What is it?
  - Primary orbital melanomas arise from melanocytes (congenital ocular melanosis, oculodermal melanosis or blue nevus)

- What Does It Look Like?
  - Progressive painful ptosis, visual blurring and scotomatas
Lymphoma

- **What Is It?**
  - Non-Hodgkin lymphoma of the orbit
  - May be associated with MALT lymphoma and Chlamydia psittaci infection (usually the result of exposure to infected birds and household pets)

- **What Does It Look Like?**
  - Typically superolateral painless mass causing palpable mass, ptosis, diplopia and abnormal ocular movement

- **Who Gets It?**
  - Caucasians, middle age

- **Treatment**
  - Diagnosis with biopsy and immunophenotyping
  - Determine primary status with full body imaging and pathologic characteristics
  - Exenteration +/- radiation and chemotherapy

Pleomorphic Adenocarcinoma

- **What Is It?**
  - Malignant transformation of a pleomorphic adenoma (either spontaneously or after incomplete excision)

- **What Does It Look Like?**
  - Similar to pleomorphic adenoma

- **Who Gets It?**
  - 60-70 years old (10-20 years older than those with pleomorphic adenoma)

- **Treatment**
  - Chlamydia psittaci infection- antibiotic therapy reduce the size of the tumor or possible cause remission
  - Surgical biopsy / resection, radiotherapy and chemotherapy are all used in various combinations
  - 65% 5-year relapse-free rate
  - Systemic dissemination is only seen in 5-10% of cases

Adenoid Cystic Carcinoma

- **What Is It?**
  - Painful rapidly growing mass in lacrimal gland

- **What Does It Look Like?**
  - CT: bony erosion, bone destruction and soft-tissue calcification

- **Who Gets It?**
  - 60-70 years old (30-20 years older than those with pleomorphic adenoma)

- **Treatment**
  - Surgical excision - lateral rhinotomy and medial maxillectomy

- **Prognosis:** survival rate correlates with the size, type, and histologic grade
  - Undifferentiated carcinoma had the worst survival rate (30%)
  - Polymorphous low-grade adenocarcinoma has the highest survival rate (96%)
Who Gets It?
- M:F
- Younger age than other lacrimal malignancies (average age is 41)

Treatment
- Radical exenteration + radiation

Prognosis poor
- 50% recurrence within 2 years
- 50% mortality rate within 1.5 years

The End!
(For those of you still awake, any questions?)