The Eye in Neoplastic Disease

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Course Goal

To provide the participant with useful clinical information about neoplastic disease, with an emphasis on ophthalmic complications.

The eye is the only part of the body where neurological and vascular tissues can be viewed directly.

Assessing lesions - ABCDE Rule

- **A** – Asymmetry (symmetry is good, asymmetry is bad)
- **B** – irregular borders (irregular borders consistent with malignancy)
- **C** – color (variation of color within lesion consistent with malignancy)
- **D** – diameter (bigger is not better)
- **E** - elevation (greater elevation consistent with malignancy)

Misc: feeder vessels as sign of malignancy

Capillary Hemangioma

- Common, benign orbital tumor
- Appears as strawberry nevus if located in more external orbit or as a blue eyelid mass if deeper in orbit
- Occurs in first year of life and slowly involutes by age 5
- Eyelid involvement may cause ptosis (lid droop) and occlusion amblyopia (lazy eye)
- Signs and symptoms:
  - Strawberry or blue mass on lid or within orbit
  - Swelling (lid) or proptosis (orbital) with crying
  - Mass may or may not be observable depending on location – orbital vs. external
- Management:
  - Orbital CT scan to determine extent of orbital involvement
  - No treatment if not affecting vision or involving the globe. Treat when lesion induces refractive error, strabismus, occlusion of vision or invasion of globe.
  - Local injection of steroids, systemic steroids, interferon, laser therapy, radiation therapy or surgical excision.
Dermoid Cyst

- Common, benign lesion
- Painless, palpable, flesh colored mass
- Composed of connective tissue
- Presents in early childhood and may slowly enlarge
- Management:
  - Orbital CT scan
  - Surgical excision by oculoplastics surgeon – cyst should be removed
  - without rupturing capsule to prevent inflammation

Rhabdomyosarcoma

- Most common primary pediatric orbital malignancy
- Most common soft tissue malignancy
- Usually occur in patients <15 years of age (usually between 7-8)

Signs and symptoms:
- Very rapid onset of unilateral proptosis
- Lid edema
- Possible lid discoloration

Management:
- CT Scan or orbit (may show bony orbital destruction)
- R/O infection (orbital cellulitis)
- Biopsy
- Systemic evaluation
- Treatment with local radiotherapy and/or systemic chemotherapy

Retinoblastoma

- Most common intraocular malignancy in children
- Tumor of the sensory retina
- 1/20,000 live births
- Most diagnosed by 5 years of age (mean age of diagnosis is 18 months)
- Most unilateral (75%), but may be bilateral
- Hereditary – but sporadic presentation possible

- Prognosis for long term survival good (85-90%)
- Prognosis for eye guarded
- Highly malignant and metastatic
- 25-30% children may develop a secondary malignancy
Symptoms:
- Strabismus (eye misalignment)
- Decreased vision
- Swelling
- Asymptomatic
- Parents may notice an unusual reflex in photos

Diagnostic signs:
- Leukocoria (white reflex) in 50-60% patients and may see vasculature in reflex
- Intraocular inflammation (due to exophytic growth)-tumor may “seed” and extend into the vitreous

Management:
- Children with a white reflex need to be referred immediately
- Examination under anesthesia (EUA)
- B-Scan and CT scan to check for calcification
- Head and orbital MRI to look for extension

Oncology consultation
- Smaller tumors: cryotherapy and laser photocoagulation
- Larger tumors: enucleation, external beam radiation therapy, episcleral plaque radiotherapy
- Enucleation necessary if most of retina is involved, invasion of anterior segment or optic nerve, vitreal seeding or irreversible vision loss

One of the reason routine eye exams are recommended on children starting at the age of 6 months.

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Choroidal Nevus

Symptoms:
- Most asymptomatic
- Typically flat or mildly elevated
- Secondary drusen common and reactivity of the RPE
Predictive Factors for Growth:
- tumour thickness
- tumour site
- flashes/floaters/blurred vision
- Orange pigment
- CNVM
- SRF (can also occur with nevi)

Melanoma
- May be external or intraocular
- Intraocular: Most common primary intraocular tumor in adults
- External: Most lethal primary skin tumor-but rare on eyelids
- Highly malignant and metastatic.
- Metastasis is most commonly to the liver. Also to lung, brain, skin and GI tract.
- Almost always unilateral
- More common in light skinned patients

- Intraocular:
  - Elevated, round mass in the choroid but protruding under or through the sensory retina.
  - Usually darkly pigmented, but may be amelanotic.

Symptoms:
- Most asymptomatic
- Blurred vision
- Scotoma or loss of visual field
- Photopsia

Treatment of Choroidal Melanoma
- Observation indicated in
  - elderly/infirm, lots of mets and very poor prognosis
  - small-medium
- Enucleate, exenterate: huge melanomas with secondary complications.
- Radiotherapy improves survival alone: plaque or external beam
- Transcleral Resection
- Multiple Treatment Modalities
- Local resection + Plaque Rx + Photocoagulation

- Management/Intraocular
- Refer for retinal evaluation/ocular oncology
- Small tumors (<10 mm diameter and <3 mm height) may be monitored carefully
- Medium tumors (10-16 mm diameter and 3-10 mm height) may treat with laser photocoagulation, external beam or episcleral plaque irradiation
- Large tumors (>16 mm diameter and >10 mm height) enucleation possible
- Evaluate for metastasis
Uveal Metastatic Lesions

- Most common intraocular tumor
- Number one primary site in women is the breast, in men it is the lung
- Choroid around 90%, ciliary body about 8%, iris 1-2%
- Uveal Metastases
  - Located in posterior pole (blood supply)
  - Unilateral or bilateral (unilateral 3 to 1)
  - Breast metastases most likely to be bilateral: lung unilateral
  - Single lesion or multiple lesions
  - RPE detachments

Management of Metastatic Tumors

- Metastatic lesions tend to be detected sooner because their posterior pole location leads to earlier symptoms
- Systemic work-up is critical
- Average survival time of 9 months after Dx

Differential Diagnoses of Metastatic Tumors

- Primary uveal melanomas
- Hemangiomas
- Osteomas
- Posterior scleritis
- Inflammatory disorders

Primary Uveal Tumors

- Unilateral & solitary
- Pigmented but may be amelanotic
- Relatively elevated
- Can break through Bruch’s membrane…”Collar Button”
- Rare in non-caucasions (C 19 X AA; H 5x AA)

Primary Uveal tumors

- Can metastasize, but rarely have by the time they are detected in the eye
- Systemic work-up a must, but not common to find metastases at time of diagnosis
- Most frequent site……..75%..........is the liver
- 2X risk of colon cancer compared to general population

COMS and other studies

- Five year survival rates for………..
- Primary choroidal melanoma
- Treatment side effects
- Main side effect of focal ocular treatment is………..Radiation retinopathy
Melanoma

• Another RR example
• Pearl
• Rapid shrinkage of the tumor with treatment may be bad news………indicates substantial malignant (and metastatic) potential

Conclusion

Optometrists are in a position to save vision, and even save a life!

Notes: