Tip of the iceberg

The proper diagnosis of a posterior segment ocular lesion may be challenging. This course will review distinct posterior ocular lesions seen in practice today. The course will highlight the associated underlying systemic disease and potential for mortality, as well as the proper management of the distinct lesion.

1. Introduction: The distinct ocular lesions
   • Choroidal
     o Nevus
     o Primary choroidal melanoma
     o Choroidal metastases
     o Melanocytoma
   • Retinal pigment epithelium (RPE)
     o Congenital hypertrophy of the RPE (CHPRE)
     o Ademocarcinoma of the RPE
   • Vascular
     o Choroidal hemangioma
     o Capillary hemangioma
     o Arteriovenous malformation

2. The ocular exam
   • Clinical presentation of ocular tumor
     o Retinal astrocytoma
       ▪ Yellow globular lesion
       ▪ May affect retina or ONH
       ▪ DDx
         • Retinoblastoma
           o Clinical presentation occurs before age 4
           o Associated intra-cranial tumors & secondary tumors
           o Classifications vary
             ▪ Familial vs sporadic
             ▪ Unilateral vs bilateral
             ▪ Hereditary vs non-hereditary
       • Diagnostic tests
         o CT: intra-tumor calcification
         o Ultrasonography: calcification
         o MRI of the brain to rule out intra-cranial tumors
      ▪ Typically asymptomatic & non-progressive
• Hence typically just followed

o Vascular
  ▪ Choroidal hemangioma
    • Red hue within the choroid
    • Isolated vs diffuse
  ▪ AVM
    • Artery to vein shunt
    • Associated ischemia & leakage
  ▪ Capillary hemangioma
    • Large dilated capillary with feeding artery & draining vein
    • Often associated with leakage

o Retinal pigment epithelium (RPE)
  ▪ Congenital hypertrophy of the RPE (CHPRE)
    • How to differentiate it from a nevus?
    • Single vs multiple
    • Associated lacunae & halo surrounding it
    • Flat dark lesion within the RPE
    • Unilateral vs bilateral presentation
  ▪ Adenocarcinoma of the RPE
    • Dark lesion with no lacunae
    • Pre-existing nodules

o Choroidal
  ▪ Nevus
    • Flat
    • <3DD
    • may be associated with drusen
    • green > amelanotic

  ▪ Primary choroidal melanoma
    • >5DD
    • Thicken
    • TFSOM pneumonic (Shield’s criteria)

  ▪ Choroidal metastases
    • Commonly associated shallow retinal detachment
    • Creamy, white or yellow colorization
    • May be multifocal or bilateral
    • Associated with mottling
- Optic nerve Melanocytoma
  - 40-50 yo darkly pigment patient
  - globular and very dark
  - extend from optic nerve
  - How do you differentiate it from juxta papillary choroidal melanoma?

- Diagnostic test used in optometric offices
  - Diagnostic value of genetic testing in choroidal melanoma
  - Ultrasound
    - Difference between primary choroidal melanoma and metastases
    - A-scan vs B-scan value
    - Vascular tumor characteristics
  - Value of photography
  - OCT
    - Case presentation
      - CHRPE
      - Choroidal tumor
      - Nevus
      - Choroidal hemangioma
    - Characteristics of choroidal tumor vs nevus using OCT
  - Others

3. Systemic Diseases
- Site of spread for choroidal melanoma
- Primary sites of tumor associated with choroidal metastases
- Conversion of nevus to a choroidal melanoma
- Secondary cancers associated with choroidal tumors
- Von Hippel-Lindau (VHL) disease (VHLD)
  - Diagnostic criteria
  - The relationship between a retinal capillary hemangioma and a cerebral hemangioblastoma
- Racemos
  - Facial involvement
  - AVM of the retina & brain
- Sturge weber syndrome (SWS)
  - Which choroidal hemangioma is likely associated with SWS?
  - Relationship with glaucoma
  - What is SWS?
- NF1
• Tuberous sclerosis
  o What is it?
  o Signs/symptoms
• Gardner's syndrome
  o Difference between associated multiple CHRPEs and bear track
  o APC gene
  o What is Gardner's syndrome?

4. Practical management approach
   a. Shield’s criteria of "to find a small ocular melanoma helpful hints daily"
   b. COMS and variability in management of choroidal tumors depending on location & size
   c. How should a patient with a nevus be followed & what test(s) should be perform?
   d. How to properly refer a patient with multiple CHRPEs?
   e. What test should be implemented for a patient suspected of having SWS?
   f. How is SWS 'secondary glaucoma managed?
   g. What test should be order for the patient suspected to have VHLD?

5. Clinical cases