Update on retinal tumors: the role of OCT in diagnosis and management
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Category: Treatment and Management of Ocular Disease: Posterior Segment

This course will highlight key diagnostic features of retinal tumors with an emphasis on incorporating spectral domain optical coherence tomography (SDOCT). This course will also address the advancing field of OCT technology including OCT angiography (OCTA) and its application to clinical practice.

**Objectives**
1. Understand when and how to use optical coherence tomography for retinal lesions and tumors
2. Understand diagnostic features of various common retinal tumors on OCT
3. Differentiate between concerning and benign features of lesions on OCT
4. Review new OCT technologies including OCTA and apply their use to management of retinal tumors

I. Update on Retinal Tumors:
   a. The role of OCT in diagnosis and management

II. Objectives
   a. 1) Understand when and how to use OCT for retinal lesions and tumors
   b. 2) Understand diagnostic features of various retinal tumors on OCT
   c. 3) Differentiate between concerning and benign features of lesions on OCT
   d. 4) Introduce advancements in OCT technology

III. Financial Disclosures
   a. None.

IV. OCT Background
   a. Retinal disease has been the driving force behind development and use
   b. Very useful in providing cross-sectional & 3D views of retinal and choroidal lesions

V. OCT Background
   a. Provides *submillimeter* detection of retinal tissue
   b. BIO = 50um discrimination
   c. Ultrasonography = 50-200um
   d. **EDI-OCT = 4um**
      i. Important to add to repertoire of diagnostic workup of suspicious retinal/choroidal lesions

VI. Asymptomatic Lesions
   a. Case 1: Optic Nerve Melanocytoma
      i. Elevated pigmented lesion on optic nerve
      ii. Often with peripapillary extension
      iii. Not until 1962 (Zimmerman) determined it to be a benign tumor
      iv. Prior to: treatment was enucleation
   b. Optic Nerve Melanocytoma
i. Benign by definition; BUT
ii. Malignant transformation occurs in 1-2% of cases
iii. 10-15% will show subtle enlargement
   1. Too subtle to be picked up on clinical examination alone
iv. Must be documented with serial OCT & fundus photography

c. Optic Nerve Melanocytoma
   i. Vision loss at any point must raise suspicion of malignant transformation/misdiagnosis
      1. Vision loss may be secondary to: RVO
      2. Tumor ischemic necrosis or vascular compression
   ii. Only 25% of cases are visually symptomatic
      1. Central vision if extension into fovea
      2. Field defect
         a. Typically enlarged blind spot

d. Clinical Appearance
   i. Dark brown/black lesion
   ii. Sometimes confined to the disc; may involve retina/choroid
   iii. Almost always unilateral
   iv. Can be associated with retinal hemorrhages, retinal edema, exudate, SRF or vitreous seeding

e. OCT Features
   i. Pseudopapilledema
   ii. Thickened ILM
   iii. Smooth transition from retina to tumor
   iv. Bright anterior border; dense optical shadowing
   v. Loss of internal retinal detail
   vi. If present: vitreous ‘seeds’ in posterior vitreous

f. Differential Diagnosis
   i. Must rule out juxtapapillary melanoma
   ii. Thickness greater than 2mm, subretinal fluid, associated lipofuscin
   iii. MRI/CT not helpful
   iv. FA can show demarcation
      1. Hypofluorescent in all phases unless subretinal fluid of disc edema is present

g. Baseline Evaluation
   i. Baseline SDOCT over the lesion
   ii. 21 line raster
   iii. 1 liner raster for further increased resolution
      1. Oversamples compared to macular cube
   iv. Baseline fundus photography, automated VF, ultrasonography

VII. Case 2
a. Case 2: Clinical Diagnostic Challenge
b. Uncertain diagnosis follow clinical examination
c. Differential diagnosis
   i. Choroidal melanoma
   ii. Choroidal nevus
iii. Epiretinal membrane
iv. ....others
d. In Other Words…
i. Bottom line:
   1. Melanoma or not?
   2. Urgent referral to ocular oncology…or not?
e. Features of Choroidal Melanoma
i. TFSOM-UHHD
   1. Thickness greater than 2mm
      a. COMS originally stated 1.5mm
   2. Fluid
   3. Symptoms
      a. Differ based on location-iris melanoma vs choroid
      b. Typically flashes, floaters/vision loss
      c. Eye pain/red eye secondary to tumor-related inflammation or secondary glaucoma
      d. 25% of choroidal melanoma are asymptomatic
ii. Features of Choroidal Melanoma
   1. Orange pigment
      a. Lipofuscin
         i. Dead vs dying tissue
   2. Margin within 3mm of optic disc
   3. Ultrasound Hollow
   4. Halo absent
   5. Drusen absent
      a. Retinal drusen in addition to drusen overlying tumor is NOT protective
      b. Not a sign of chronicity as retinal drusen may have been present first
f. Choroidal Melanoma Features on OCT
   i. Increased choroidal thickness
   ii. Subretinal fluid
   iii. Lipofuscin
   iv. Shaggy photoreceptors
g. Our Case: OCT Findings
   i. No subretinal fluid
   ii. Thickened internal limiting membrane (ERM)
   iii. Disorganized and thickened retinal layers with ellipsoid zone loss
h. By Definition
   i. Retina & RPE disorganization with overlying ERM
   ii. Combined hamartoma of retina and retinal pigment epithelium
i. CHRRPE
   i. Benign lesion
   ii. Vision loss only if lesion extends into the fovea
   iii. Almost always unilateral
iv. Systemic associations include NF-II, branchio-oculofacial syndrome, Gorlin-Goltz syndrome
v. Congenital
vi. Treatment considered for macular ERM
j. Prognosis
   i. Excellent; minimal progression expected

VIII. Case 3
a. Case 3: Choroidal Nevus
   i. 1/4800 choroidal nevi transform into melanoma
   ii. 6.5% of patients over 49 have a choroidal nevus
b. OCT Findings
   i. Choriocapillaris compression
   ii. Obstruction of blood flow to RPE and outer retinal layers = ellipsoid zone loss
   iii. Optical shadowing
   iv. Lesion may have subretinal fluid
      1. Alone-not an indicator of definite malignant transformation
v. TFSOM-UHHD; 3+ features = 50% chance of malignancy
c. Baseline Evaluation of Choroidal Nevi
   i. Baseline OCT & fundus photography & ultrasonography
   ii. For a ‘low risk’ lesion:
      1. Repeat in 6 weeks to 3 months; if no change:
         a. May choose to repeat in another 3 months
      2. Repeat in 6 months; if no change:
      3. Repeat annually
         a. Or sooner if new symptoms are noted
      4. Monitor more frequently based on risk factors
d. Differential Diagnosis
   i. Of course; choroidal melanoma
      1. Uveal melanoma = 85% of all ocular melanoma arise from melanocytes
         a. Mean age of Dx is 60 years old
e. Gene Expression Profiling
   i. Commercially available
   ii. Provides prognostic significance
      1. Class 1 (low metastatic risk)
      2. 1A: 2% change of metastasis over 5 years
      3. 1B: 21% chance of metastases over 5 years
      4. Class 2 (high metastatic risk)
         a. 72% chance of metastases over 5 years

IX. Symptomatic Lesions
a. Case 4: Central Serous Chorioretinopathy?
   i. Chief complaint
      1. “Blurred vision in my right eye”
      2. History of unknown IV in ED for respiratory/allergic reaction after difficulty breathing
ii. Clinical appearance
   1. Typical
   2. Opted to monitor without treatment

iii. OCT Findings
   1. Serous RPE detachment
   2. subRPE debris
      a. Not true shaggy photoreceptors, but similar appearance
   3. Near complete resolution after 7 weeks
   4. No treatment initiated

b. Case 4b: CSCR ?
   i. Patient chief complain of unilateral “blurred central vision”
      1. History of recent increased family stress
   ii. Clinical examination
      1. “Identical”
   iii. OCT Findings
      1. RPE detachment
      2. Sub RPE serous fluid
      3. Minimal improvement over time
   iv. Patient improved and worsened over a 1.5 year period
   v. Diagnosis
      1. Diffuse choroidal melanoma
         a. Patient was ultimately treated with plaque brachytherapy with ultimate improvement in subretinal fluid
      2. Other masquerades
         a. Diffuse choroidal hemangioma
         b. Sturge-Weber Syndrome
            i. Port-Wine stain with neurosensory detachment
         c. Role of oral propanolol?
      3. FA is key in the Ddx in these cases
      4. Non-resolving ‘CSCR’ must undergo further evaluation

X. Case 5
a. 61 year old white male
b. HM acuity OD
c. Reported to be stable since at least 1979
   i. No treatment was pursued or recommended
d. Choroidal Osteoma
   i. Tumor comprised of mature bone tissue
   ii. Not known to be associated with any systemic condition
   iii. No association with serum calcium, phosphorus, CBC abnormality
   iv. Typically exhibit slow growth (50%)
   v. Average of +0.37mm basal diameter per year
e. Choroidal Osteoma
   i. Decreased acuity secondary to:
1. 1) Decalcification with RPE and choriocapillaris atrophy
   2. 2) Serous retinal detachment
   3. 3) Choroidal neovascularization

XI. Continued OCT Development
   a. OCT Angiography
      i. Non-invasive imaging technique used to visualize retinal and choroidal vasculature
      ii. Multiple B-scans acquired at the same location over a period of time
      iii. B-scans differ at the same location with erythrocyte flow
      iv. Microvasculature is visualized through decorrelated B-scans
      v. Results in an image of moving erythrocytes in retinal and choroidal vasculature
   b. OCTA
      i. En face flow information and cross sectional structural information
      ii. Not a replacement for FA/OCT
      iii. Provides new information
   c. Clinical Uses
      i. Choroidal neovascularization
         1. Possibly aiding in re-treatment criteria
      ii. Diagnosis of retinal vascular disease
         1. Mactel type 2
         2. RVO
         3. Diabetic retinopathy
            a. Identifying microaneurysms
            b. Macular ischemia
            c. Confirming presence of NV
      4. AMD
      5. Non-exudative lesions in eyes with intermediate AMD
   d. Disadvantages of OCTA
      i. Static blood flow information
      ii. No leakage, pooling or staining
      iii. Small field of view
         1. 3x3mm or 6x6mm HD give highest resolution
         2. 6x6mm and 8x8mm have reduced resolution
   e. Swept-Source OCT
      i. Ability to provide wide-field imaging
      ii. Longer wavelength than SD-OCT
      iii. Ability to image deeper structures
      iv. Decreases effect of media opacities
   f. Advancements in General
      i. Higher speeds
      ii. Increased resolution
      iii. Wider field
   g. **Goal is for subtle retinal, choroidal and vitreal changes to be more easily measured**
h. Pearls
   i. One line raster over samples by 20x vs macular cube
   ii. EDI reduces resolution of vitreomacular interface
      1. Can perform ‘reduced’ depth imaging with 1 line raster
         offset anteriorly
         a. Excellent for VMI disorders
   iii. Finally
      1. Always scan BOTH eyes

XII. Summary
   a. OCT is very useful to aid in diagnosis of unknown retinal tumors and
      lesions
   b. OCT can aid in determination of necessity (and/or timeliness of)
      retinal/oncology referrals
   c. OCT is an important adjunct to clinical examination and other ancillary
      tests in monitoring retinal tumors
   d. Continued advancements aim to increase diagnostic ability of choroidal,
      and retinal lesions

XIII. Thank you
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