Title: Over the Hill: Type II Juxtafoveal Telangiectasia with Secondary Cystoid Macular Edema

Abstract: This case features a patient with type II juxtafoveal telangiectasia with subsequent cystoid macular edema. The outline details case information, differential diagnoses, stages of the macular telangiectasia, and diagnostic and management strategies.

I. Case History
   • Demographics: 83-year-old Caucasian male
   • Chief complaint: Mild-to-moderate decrease in vision OS worsening over the past two months
   • Ocular History: Pseudophakia, OU
   • Medical History: Hypertension, hyperlipidemia, vertigo, chronic obstructive pulmonary disease, colonic polyps, osteoarthritis, restless leg syndrome, gout, peripheral vascular disease, and allergic rhinitis
   • Medications: Memantine, flunisolide, rivastigmine tartrate, omeprazole, atorvastatin calcium, meloxicam, losartan potassium, tramadol hydrochloride, cetirizine, docusate sodium, chondroitin/glucosamine, acetaminophen, aspirin, multivitamin

II. Pertinent Findings
   • Entrance Testing
     • Best Corrected Visual Acuities – OD: 20/20-2, OS: 20/25-2, OU:PHNI
     • Pupils – 3mm, equal, round and reactive to light, OD & OS, no relative afferent pupillary defect
     • Tonometry – OD: 11mmHg, OS: 12mmHg
     • Extraocular Muscle Movements – full/no restrictions OD & OS
     • Confrontations – full to finger counting OD & OS
     • Amsler Grid – OD: normal, OS: small central area of metamorphopsia
   • Slit Lamp Evaluation
     • Cornea – clear corneal incision OD & OS
     • Conjunctiva – clear OD & OS
     • Anterior Chamber – deep & quiet, (-) cells or flare OD & OS
     • Iris – normal OD & OS
     • Lens – posterior chamber intraocular lens, clear and centered, capsule intact, OD & OS
   • Posterior Segment Findings
     • OD: Juxtafoveal telangiectatic vessels, microaneurysms in macula, reticular degeneration in periphery; all other findings within normal limits
       • Spectral-domain optical coherence tomography (SD-OCT) performed shows central RPE changes with mild disruption of the ellipsoid zone, no intraretinal or subretinal fluid, normal foveal contour, no neovascularization
- Fundus autofluorescence (FAF) photo – increased hyperautofluorescent signal at the fovea
- Fluorescein angiography (FANG) – juxtafoveal telangiectatic vessels that hyperfluoresce early and show late phase leakage
- OS: Juxtafoveal telangiectatic vessels, microaneurysms in macula, mild macular elevation, pavingstone degeneration inferiorly; all other findings within normal limits
- SD-OCT performed shows central RPE changes with mild disruption of the ellipsoid zone, intraretinal cystic cavitations with retinal elevation, disrupted foveal contour, no neovascularization
- FAF photo – increased hyperautofluorescent signal at the fovea
- FANG – juxtafoveal telangiectatic vessels that hyperfluoresce early and show late phase leakage, hyperfluorescence in late stage greater OS than OD and in an expansile dot appearance

III. Differential Diagnoses
- Cystoid macular edema secondary to macular telangiectasia
- Diabetic macular edema
- Epiretinal membrane
- Irvine-Gass syndrome

IV. Diagnosis and Discussion
- Case Diagnosis: Presumed type II idiopathic juxtafoveal telangiectasia (IJT), OU with cystoid macular edema (CME), OS.
  - Diabetic macular edema ruled out due to lab-proven absence of diagnosis
    - Last HbA1c was 5.6 and glucose ranges were below 100 per patient’s primary care physician/lab testing
  - Irvine-Gass syndrome ruled out due to uncomplicated cataract extractions performed six years ago without history of post-operative CME
  - Etiology confirmed by retinal specialist at Ann Arbor VAMC
- Secondary Diagnoses: Pseudophakia, OU
- Diagnostic Techniques:
  - Fundoscopic findings of IJT type II include reduction of retinal transparency, crystalline deposits, ectatic capillaries, retinal pigment plaques, right-angle vessels, foveal atrophy and neovascular complexes
  - One of the earliest changes seen in IJT type II is an increased short-wavelength FAF signal in the foveal region due to loss of macular pigment
  - FANG is the gold standard for diagnosis of IJT type II
  - Angiographic areas of leakage do not correlate with the presence of CME or retinal thickening on OCT
  - Research shows confocal blue reflectance (CBR) imaging may be almost as sensitive as FANG to detect IJT type II
- Discussion:
  - Idiopathic Juxtafoveal Telangiectasia (IJT) type II is the most common type, occurring bilaterally in middle-aged women and men
  - Pathogenesis is unclear, but evidence points to parafoveal Mueller cell dysfunction as opposed to a primary abnormality of the retinal capillaries
    - Perifoveal depletion of Muller cells has been seen on histopathology
• Retinal crystalline deposits are thought to represent the footplates of degenerated Muller cells
• Presentation shows occult non-exudative IJT, which is difficult to detect on ophthalmoscopy; however, has diagnostic features on fluorescein angiography and optical coherence tomography
• A classification system by Gass denotes the five distinct stages of IJT type II
  • Stage 1: diffuse juxtafoveolar hyperfluorescence in late-phase FANG
  • Stage 2: reduced parafoveolar retinal transparency
  • Stage 3: dilated right-angled venules
  • Stage 4: intraretinal pigment plaques
  • Stage 5: secondary neovascular membranes
• Long-term prognosis of IJT type II may be poor when associated with persistent macular edema, central retinal atrophy, macular hole or subretinal neovascularization (SRNV)
  • SRNV in IJT type II appears to originate from the deep retinal plexus with a predilection site temporal to foveal center
    • Unlike choroidal neovascularization in age-related macular degeneration (AMD), SRNV in IJT type II is not usually accompanied by a RPE detachment
    • SRNV is usually smaller and less aggressive in comparison to AMD

V. Treatment and Management
• Extensive patient education on findings and signs/symptoms of progressive macular edema
  • Patient advised to monitor with home Amsler grid
• Close observation of CME is indicated as no treatment has been proven successful in cases of CME secondary to IJT
  • Monitor every two months with dilated fundus exam and SD-OCT
• Currently there are also no effective treatment options for non-proliferative IJT type II
  • Argon laser photocoagulation, photodynamic therapy, intravitreal steroid injections and intravitreal injections of vascular endothelial growth factor (VEGF) inhibitors have all been tested without long-term success
• Research shows benefit from intravitreal anti-VEGF injections for proliferative stage
  • Insufficient evidence in favor of a specific treatment regimen
• IJT type II is a primary neuroretinal degenerative disease with secondary retinal vascular involvement
  • Long-term effectiveness of treatments that target altered retinal capillaries may be limited
  • Neuroprotection may play a significant role in future treatment
  • Some researchers believe blockage of VEGF, with its inherent neuroprotective properties, might even accelerate the disease course
  • The first clinical trial following a neuroprotective strategy is underway
  • Current research shows that supplementation with lutein, meso-zeaxanthin and zeaxanthin may stabilize vision and improve the cavitations in this condition

VI. Conclusion
• New imaging tools such as OCT angiography and FAF provide insights into the diagnosis and progression of the disease
• IJT is a primary neurodegenerative macular disease with secondary vascular involvement
• Current research with neuroprotective treatment strategies are underway
• Currently there are no successful treatment options for IJT without neovascularization

References:


Chen JJ, McAllister, AR, Sohn EH. Idiopathic Juxtafoveal Telangiectasia Type II (Macular Telangiectasia type 2). EyeRounds.org. February 17, 2014; Available from: http://eyerounds.org/cases/185-JXT.htm


