Abstract
Outer retinal tubulation (ORT) represents photoreceptor rearrangement in degenerative outer retinal diseases (Zweifel). It is important to differentiate ORTs because they do not indicate an active neovascular, exudative process and thus do not require treatment (Iaculli).

I. Case History
   a. Demographic:
      i. 83 year old, Caucasian, male
   b. Reason for Visit:
      i. Referred from a community clinic for exudative, age-related macular degeneration
   c. Chief Complaint:
      i. Reported good vision with current single vision distance and near spectacles
   d. Ocular History:
      i. Motorcycle accident in the 1990s with unknown damage to the left eye
   e. Medical History:
      i. Hearing loss, hypertension, hyperlipidemia

II. Pertinent Findings
   a. Initial Exam
      i. Best Corrected Visual Acuity
         1. OD: 20/30-, 0.4/0.6M, OS: 20/40-, 0.4/1.0M
      ii. Anterior Segment
         1. Cornea:
            a. OU: clear, mild arcus 360
         2. Lens:
            a. OU: 1+ anterior cortical changes, nuclear sclerosis
         3. Vitreous:
            a. OU: posterior vitreous detachment with Weiss ring present
      iii. Posterior Segment
         1. Optic Nerve:
            a. OU: 0.30 round, shallow, borders distinct, healthy
         2. Macula
            a. OD: scattered small to intermediate size soft drusen, with mild epiretinal membrane
            b. OS: scattered small to intermediate size soft drusen, geographic atrophy superior to fovea (1 disc diameter vertical/1.5 disc diameter horizontal)
      iv. Optical Coherence Tomography (OCT) Findings
         1. OD: multiple small-intermediate size soft drusen with disrupted ellipsoid zone, parafoveal epiretinal membrane traction
         2. OS: large geographic atrophy superior to fovea, several round lesions with a hyporeflective interior surrounded by a hyper-reflective border located in the outer nuclear layer of the retina, disrupted ellipsoid zone
   b. Follow-Up Visit (1 year later)
      i. Best corrected visual acuity
         1. OD: 20/40-2, OS: 20/50-1
ii. Optical Coherence Tomography (OCT) Findings
   1. OD: multiple small-intermediate size soft drusen with disrupted ellipsoid zone, parafoveal epiretinal membrane traction, overall stable to 2014 scan
   2. OS: large geographic atrophy superior to fovea, several round lesions with a hyporeflective interior surrounded by a hyper-reflective border located in the outer nuclear layer of the retina, Raster scans show stability in size of some lesions and decreasing size in other lesions, total number of lesions stable to 2014 scan

III. Differentials
   a. Exudative, age-related macular degeneration
      i. Shared Signs: hyporeflective optical coherence tomography findings indicate fluid, disrupted ellipsoid zone, presence of soft drusen
      ii. Differentiating Signs: typically shallower and diffuse or dome-shaped without a hyper-reflective border surrounding area of fluid
   b. Central serous chorioretinopathy
      i. Shared Signs: hyporeflective optical coherence tomography findings indicate fluid
      ii. Differentiating Signs: typically large dome-shaped pigment epithelial detachment with intact outer retinal layers
   c. Macular edema
      i. Shared Signs: hyporeflective optical coherence tomography findings in retinal layers
      ii. Differentiating Signs: round cystic spaces without a hyper-reflective border usually found in inner retinal layers, no hemorrhages, no exudates on slit lamp examination

IV. Diagnosis and Discussion
   a. Diagnosis: outer retinal tabulation
      i. Distinguishing Features: hyper-reflective border surrounding hypo-reflective interior, found in the outer retinal layers along the border of healthy retina and areas of absent or degenerating retinal pigment epithelium
   b. Discussion:
      i. First described by Zweifel, commonly found in age-related macular degeneration and other outer retinal diseases. Imaged for the first time due to advances made by spectral-domain optical coherence tomography.
      ii. Not limited to age-related macular degeneration, can also be found in other outer retinal conditions such as multifocal chroiditis, Best’s disease, and Bietti crystalline dystrophy (Goldberg, Iriyama).
      iii. Associated with subretinal fibrosis, especially those with classic choroidal neovascularization, and worse visual acuity at baseline (Faria-Correria).
      iv. Hyper-reflective border is a combination of the external limiting membrane and inner segment mitochondria. Lumen contains degenerating photoreceptors and acts as a potential space that can be filled with fluid. Histology studies show outer retinal tubulation formation occurring in four distinct stages. Initial disruption of the interdigitations between outer segment photoreceptors and the retinal pigment epithelium by subretinal fluid starts the process (Schaal, Litts).
      v. Found commonly along the borders of geography atrophy and healthy outer retinal tissue, associated with worse visual acuity at baseline, larger area of choroidal neovascularization lesion and geographic atrophy (Lee).
vi. Enlargement of geographic atrophy is greater in eyes without outer retinal tubulation (Hariri).

V. Treatment and Management
   a. In studies, outer retinal tubulation remained stable after treatment with intravitreal ranibizumab or bevacizumab for exudative macular degeneration but did show temporary decrease in size of tubules (Zweifel, Iaculli, Goldberg, Dirani)
   b. Long-term follow-up of a patient with exudative, macular degeneration and treated with intravitreal ranibizumab for three years also showed continued presence but changes in size (Jung)
   c. Outer retinal tubulation seen in 30% of patients treated with ranibuzimab and continued incidence despite visually effective treatment. Also, patients with outer retinal tubulation present before treatment benefitted less with treatment (Dirani)

VI. Conclusion
   a. With increasing use of spectral domain optical coherence tomography in patient management, it is essential to correctly identify ocular conditions which warrant further follow-up and treatment.
   b. Outer retinal tubulation (ORT) is a degenerative process which appears secondary to other disease states that affect the outer retinal layers such as age-related macular degeneration.
   c. Currently, there are no treatments for this condition, but it is important to monitor the changes of the primary disease that caused outer retinal tubulation formation.
   d. Outer retinal tubulation resembles other retinal conditions such as macular edema or choroidal neovascularization and correct identification will prevent over-referral and over-treatment.
Bibliography


Hariri A, Nittala MG, Sadda SR. Outer retinal tubulation as a predictor of the enlargement amount of geographic atrophy in age-related macular degeneration. Ophthalmology 2015; 122: 407-413.
