Torpedo Maculopathy: Proper Identification and Management of an Unusual Macular Lesion

I. Case History

- **Patient demographics**: 68 year old white male, presents for CEE
- **Chief complaint**: ‘Readers seem too strong’ and he would like an adjustment to his NVO glasses.
- **Ocular history**:
  - Type II DM x 12 years, h/o mild non-proliferative diabetic retinopathy without CSME OU
  - *managed with insulin and metformin
  - *Last A1c 8.0% (4/19/16)
  - Scar adjacent to macula OS; stable and asymptomatic per previous note
  - Dry eye syndrome OU, patient is symptomatic and uses artificial tears
  - S/p bilateral blepharoplasty (9/2015), patient is very happy with the results
  - Refractive error OU
- **Medical History**: cough, obstructive sleep apnea, dermatitis, diabetes mellitus type II, restless leg syndrome, abdominal pain, anxiety, personal history of colon polyps and alcoholism, hypertension, gastroesophageal reflux disease, ischemic heart disease, s/p knee/elbow/neck surgery, diverticulitis, umbilical hernia, fatigue, cardiomyopathy
- **Medications**: accu-check, aspirin, atorvastatin, baclofen, cetirizine HCL, clopidogrel bisulfate, cyanocobalamin, cyclobenzaprine, hydrochlorothiazide, insulin (Novolin), lisinopril, metformin, metoprolol succinate, omeprazole, pramipexole dihydrochloride, saxagliptin, sertraline HCL, sodium fluoride, spironolactone, temazepam
- **Allergies**: intravascular contrast media, Dilaudid (hydromorphone), ciprofloxacin
- **Other salient information**:
  - (-) flashes, floaters, eye pain, nor trauma to either eye
  - (-)FHx: glaucoma, blindness, AMD
  - Patient grew up in the greater-Boston area with no farm exposure. Never lived in Ohio-Mississippi River Valley

II. Pertinent findings

- **Clinical**:
  - **DVAcc**: OD: 20/25, OS: 20/20, NVAcc: 20/25 OU
  - **Pupils**: ERRL, (-)APD, **EOMs**: SAFE, no diplopia/pain, **FCF**: FTFC OD, OS, **Color Vision**: 11/11 OD, OS, **Facial Fields**: Full OD, OS
  - **Refraction**: OD: +2.00sph, 20/20 OS: +1.00sph, 20/20 **Add**: +1.50 20/20 OU (longer working distance)
  - **SLE**: L/L: Clear OU without dermatochalasis OD,OS
  - Conj: W&Q OU
  - Cornea: Clear OD, OS, no evidence of FB/scar OU
  - AC: Deep and Quiet OU
  - Iris: Flat and clear, (-)NV1 OU
  - VH: 1:1 N&T OU
  - **IOP**: 16/16 OD/OS GAT, Time: 9:35am
  - **DFE**: Lens: tr NS, OD, OS
  - Media: clear OU
  - Margins: Distinct OU
  - C/D: 0.30r OD, OS
  - Rim: pink and healthy OD, OS
  - Macula: OD: clear, no evidence of CSME
OS: oval area of hypopigmentation with hyperpigmentation/RPE hyperplasia at temporal edge. Oval appears temporal from fovea, not involving fovea, no evidence of CSME

Vessels: regular OU
Background: scattered H/Ma's <photo2A OD, OS
Periphery: Retinae intact 360 degrees
- **Fundus photos** were performed OD, OS
- **OCT:** posterior pole and macula OD, OS, EDI scans and AF features performed as well
  - OD: normal foveal contour, (-)SRF/IRF
  - **OS:** normal foveal contour, choroidal excavation sup-temp to fovea, RPE intact, photoreceptor layer potentially atrophied, outer retinal layers are less distinct within area of excavation, (-)IRF. Size of lesion: V:1.598mmxH:2.627mm (1x1.6DD) consistent with torpedo maculopathy type II

- **HVF 24-2:**
  - OD: clean field
  - OS: central inferior-nasal defect consistent with location of torpedo lesion; defect appears absolute

- **Physical:**
  - Good facial symmetry, no ptosis OU

- **Laboratory studies:**
  - Last A1c: 8.0% (4/19/16)
  - Last BP: 117/74 (7/14/16)

- **Radiology studies:** N/A

- **Others:** N/A

**III. Differential diagnosis**
- **Primary/leading:** Torpedo Maculopathy OS
- **Others:** Toxoplasmosis scar, CHRPE, Histoplasmosis spot, Amelanotic nevus, Traumatic scar

**IV. Diagnosis and discussion**
- **Elaborate on the condition:** Torpedo maculopathy Type II
  - Torpedo maculopathy is a rare, congenital condition that presents unilaterally. The lesion is flat, oval shaped and has well-defined borders.
  - The lesion was first described in 1992 by Roseman and Gass, “hypopigmented nevus of the retinal pigment epithelium” (Roseman and Gass, 1992); shortly after it was renamed torpedo maculopathy. The lesion is occasionally referred to as ‘solitary hypopigmented nevus’
  - Although the lesion can result in a visual field defect, it rarely affects central visual acuity (Golchet et al.)

- **Expound on unique features:**
  - Unilateral, flat, oval shaped ( torpedo shaped) area of hypopigmentation temporal to the fovea with RPE migration/hyperplasia at the temporal edge of the lesion. The lesion is usually longer horizontally than it is vertically
  - There are two types of torpedo maculopathy, type I and type II. Type I versus type II torpedo maculopathy are differentiated using the OCT: type I demonstrates outer retinal structure attenuation without retinal or choroidal cavitation, while type II has outer retinal structure attenuation and retinal and/or choroidal cavitation. (Wong et al., 2014). Our patient has OCT findings consistent with torpedo maculopathy type II
  - Retinal and choroidal excavation often remains stable (Wong et al.), but few cases have identified sub-retinal fluid in association with torpedo maculopathy. (Trevino et al.) This is identified as elevated retinal tissue via OCT; our patient lacked this feature.
  - Visual field defects are variable, but all subjects with type II torpedo maculopathy in the Wong et al. study had some sort of visual field defect, this is also consistent with our patient
  - While this lesion is congenital, the etiology of the lesion is often debated. An article in 2010 provided a reasonable etiology, suggesting that the lesion derived from the ‘site of the fetal bulge’
A ‘bulge’ forms in the temporal-macular retina during 4-6 months gestation, and while this bulge retracts during later months of gestation, it is thought that a mild residual depression remains. It is postulated that a disruption in the RPE occurs at this time, resulting in the torpedo lesion.

- Torpedo maculopathy versus other similar lesions:

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Appearance</th>
<th>Location</th>
<th>OCT Features</th>
<th>Visual Field Features</th>
<th>Laterality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Torpedo Maculopathy</td>
<td>Flat, oval hypopigmented with RPE hyperplasia at temporal edge</td>
<td>Adjacent/temporal to fovea</td>
<td>Type I: outer retinal attenuation Type II: outer retinal attenuation and retinal and/or choroidal excavation</td>
<td>Visual field defect present in type II, variable in type I</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Toxoplasmosis Scar</td>
<td>Round or oval with pockets of hypopigmentation and RPE hyperplasia</td>
<td>Variable, can present throughout macula</td>
<td>Retinal atrophy and inner and outer retinal layer disruption</td>
<td>Absolute defect</td>
<td>Unilateral</td>
</tr>
<tr>
<td>CHRPE</td>
<td>Round with dense RPE pigmentation and pockets of hypopigmentation (lacunae)</td>
<td>Variable, often mid-periphery; rarely in the macula</td>
<td>Thickened RPE, absence of RPE within lacunae. Outer retinal loss and occasional subretinal cleft</td>
<td>Defects present</td>
<td>Variable</td>
</tr>
<tr>
<td>Histoplasmosis</td>
<td>Multiple focal areas of hypopigmentation, PPA around ONH</td>
<td>Posterior pole; rarely in mid-periphery (5% of patients)</td>
<td>Outer retinal atrophy and outer retinal band disruption</td>
<td>Defects present</td>
<td>Variable</td>
</tr>
<tr>
<td>Amelanotic Nevus</td>
<td>Round hypopigmented lesion</td>
<td>Variable, often mid-periphery and posterior pole; rarely in the macula</td>
<td>Unclear</td>
<td>Unclear</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Traumatic Scar</td>
<td>Variable shape with areas of RPE hyperplasia and atrophy</td>
<td>Variable</td>
<td>Disorganization of RPE and choriocapillaris; variable disruption of inner and outer retina</td>
<td>Defects present</td>
<td>Unilateral</td>
</tr>
</tbody>
</table>

(Fung et al., Kolomeyer et al., Lee et al., Monnet et al., Oliver A et al., Villegas et al., Wong et al.)

V. Treatment, management

- Treatment and response to treatment:
  - Observation is appropriate and baseline visual fields are recommended; annual DFE (Teitelbaum et al.)
  - Many case study authors have described torpedo maculopathy as non-progressive, but Wong et al. postulate that the lesions may progress very slowly over many years (Shields et al); however, no treatment/intervention is necessary for retinal/choroidal excavation (Wong et al., 2014)
  - Although this patient has diabetes and mild non-proliferative retinopathy, the torpedo lesion can be differentiated from diabetic macular edema due to the retinal layers involved, and lack of increased macular thickness. Macular thickening warrants a retinal consult
- Refer to research where appropriate: Article authors included next to statements to support findings
- Bibliography, literature review encouraged:

VI. Conclusion
- Clinical pearls, take away points if indicated:
  - Our patient’s unusual macular lesion can be properly identified as torpedo maculopathy using funduscopic examination
  - The lesion can be further classified as torpedo maculopathy type II using optical coherence tomography (OCT)
  - Choroidal and retinal excavation as identified with OCT can be monitored with observation and do not require retina management/intervention. If retinal elevation/thickening are present, then a retinal consult is warranted
  - A baseline field should be obtained for future comparison
  - These cases can become more difficult to manage in the presence of other disease etiology, such as diabetes or macular degeneration. It is important to correctly identify the lesion in order to determine the prognosis and proper management. Funduscopic photography, OCT and visual field are useful tools and will aid the diagnosis and management