Moderate Vision Impairment with a Normal Appearing Fundus
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A 61-year-old white male presents with progressive bilateral decreased vision, normal fundoscopy, and no family history. Multifocal ERG confirms diagnosis of occult maculopathy. Patient is subsequently referred to low vision clinic to address functional needs.

I. Case History
a) Patient demographics: 61 year old white male
b) Chief complaint: Gradual decrease in distance vision in the left eye more than the right eye, starting about ten years ago. It has become more concerning in the last five years due to difficulty seeing exit signs when driving.

b) Ocular Hx:
   - “Corneal dystrophy”

c) Medical Hx:
   - Hypertension
   - Arthritis
   - Pacemaker due to h/o aneurysm in ascending aorta

d) Medications:
   - Carvedilol
   - Indomethacin
   - Losartan
   - Meloxicam
   - Omeprazole

II. Pertinent findings
a) Clinical
   BCVA: OD: 20/60
       OS: 20/70
   Pupils: PERRL, no APD
   EOMs: Full
   Color Testing (Farnsworth D15): WNL OD and OS
   Contrast (Mars): OD: 1.36 log
                    OS: 1.28 log
                    OU: 1.44 log

   Anterior Segment:
   Lids/Lashes: mild MGD OU
   Conj: white and quiet OU
   Cornea: mild arcus OU, mild mapdot dystrophy superiorly OD>OS
   Angle: 1:1 OU
   AC: deep and quiet OU
   Iris: flat OU
   Lens: trace NS OU
IOP: WNL OU
Posterior Segment:
  Vitreous: Syneresis OU
  Macula: flat, even OU; no foveal reflex OU
  Posterior Pole: WNL OU
  ONH C/D: OD: 0.10, no pallor
  OS: 0.05, malinserted, no pallor
Periphery: clear to ora 360

b) Other Testing
  • Cranial Nerve Testing: normal
  • Goldmann Fields: I-4-e: full OD and OS
  V-4-e: full OD and OS
  • OCT: disruption of IS/OS junction subfoveally OD and OS
  • MRI with and without contrast: unremarkable imaging of the brain,
    optic nerves, and globe
  • Full field ERG: near normal rod and cone responses with normal
    implicit times OU
  • Multifocal ERG: reduced central ring ratio amplitudes consistent with
    maculopathy OU

III. Differential diagnosis
  • Occult maculopathy (Primary)
  • Functional vision loss
  • Age Related Macular Degeneration
  • Chloroquine Retinopathy
  • Stargardt’s Macular Dystrophy
  • White Dot Syndrome (Acute Zonal Occult Outer Retinopathy)
  • Optic Neuropathy

IV. Diagnosis and discussion
  Occult maculopathy is an inherited or sporadic disease localized to the
  photoreceptors in the macula. The age of onset for progressive bilateral vision loss is
  typically 31-60 years old. The vision loss is usually bilateral, but may be asymmetric. In
  most cases, visual acuity remains 20/200 or better in at least one eye. On clinical
  examination, normal appearing fundoscopy and normal fluorescein angiography may lead
  to a misdiagnosis of functional vision loss. Further testing with a normal full field ERG and
  reduced central amplitudes on multifocal ERG is pathognomonic for occult maculopathy.
  More recent literature has found abnormal thinning of the foveal thickness that is
  associated with the disruption of the IS/OS line and COST line on SD-OCT. Many patients
  may also show an abnormal IR reflectance around the fovea, corresponding to the area of
  IS/OS disruption. Fundus autofluorescence show no abnormalities, indicating normal
  retinal pigment epithelium.
  Unique features of this case pertain to the patient’s initial diagnosis of
  unexplained vision loss. With subtle changes on OCT and an extensive work-up with full-
  field ERG and multifocal ERG, patient was diagnosed with occult maculopathy and
subsequently referred to the low vision clinic. As a fairly healthy 61 year old with mostly normal ocular health other than the localized cone dystrophy, patient presents as an ideal candidate for multiple low vision devices, including a bioptic telescope.

V. Treatment, management
   a) Treatment: Low vision aids
      • 2.2x bioptic telescope for mobility and driving was prescribed. It is an assistive device to wear while driving for persons with moderate central vision impairment, ranging from 20/40 to 20/100 or 20/200 (depending on the state DMV standards). The bioptic is mounted in the upper portion of a regular spectacle lens (“carrier” lens) and the driver views through the carrier lens for the majority of the time, dipping the head very briefly to spot signs as needed.
      • 2.5x monocular telescope for distance spotting
      • High plus prism readers were prescribed for reading
      • 3.5x/8D illuminated pocket magnifier (Easypocket) was provided for spot reading
      • Gray fitover medical filters for glare/photosensitivity
   b) Bibliography

VI. Conclusion
   Progressive, bilateral vision loss with normal fundoscopy and normal fluorescein angiography may be an indication for a more detailed look at the IS/OS line and foveal thickness on SD-OCT to rule out occult maculopathy. Further testing with a normal full-field ERG and an abnormal multifocal ERG will confirm the diagnosis. Patients with occult
maculopathy are managed with referrals to low vision for functional aids. In this case, patient was fit with telescopes to address distance difficulties, stronger reading glasses and a pocket magnifier for near, and a medical filter for photosensitivity.