Differential Diagnosis and Management of Systemic Lupus Associated Bilateral Optic Neuropathy

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Abstract: This case report outlines possible ocular manifestations associated with systemic lupus and shows how differential diagnosis and ancillary testing play an important role in critical thinking and clinical judgment.

Case Report:

- **Case History:**
  - **Demographics:** 73 y/o African American Female
  - **Chief Complaint:** Painless, blurry vision OU worsening x 4 years, worse with nighttime driving
  - **Ocular History:** H/O toxic maculopathy associated with prior Plaquinil use, cataracts, dry eye, (-) ocular surgeries
  - **Medical History:** Systemic lupus, GERD, arthritis, anemia, sleep apnea, anxiety and Raynaud's phenomenon. FHx of HTN, DM, arthritis and thyroid disease.
  - **Medications:** (-) ophthalmic medications. Systemic medications include: Azathioprine, Clopidogrel, Fluoxetine, Gapapentin, Triamterene, Omeprazole, Prednisone and Zolpidem.

- **Pertinent Findings:**
  - **Clinical:**
    - DVA sc: 20/50 OD, 20/30 OS
    - Super Pinhole Acuity: 20/20 OD, 20/20 OS
    - Brightness Acuity Test: 20/40 OD, 20/40 OS.
    - IOP 15mmHg OD/OS.
    - SLE-Central vacuoles, trace cortical opacification and 2+ nuclear sclerosis cataract formation OD/OS.
    - DFE- enlarged C/D ratio OU (0.6 OD/OS), trace ERM OD/OS with macular pigment mottling OD/OS
  - **Physical:** The patient displayed classic signs of systemic lupus (thinning of the hair, joint pain (used a cane for walking support), and malar facial rash across the cheeks of the face.)
  - **Laboratory studies:**
    - ONH OCT w/ significant RNFL loss, OD>OS
    - Macular OCT w/ significant thinning of the maculas OU
    - At future FU exams- HVF 24-2 OU, fluorescein angiography OU and fundus autofluorescence OU were ordered which revealed a central defect OU, delayed transit time OU and significant retinal thinning along the temporal arcades OU.

- **Differential Diagnosis:**
  - Primary/leading Diagnosis:
    - Lupus Associated Optic Neuropathy
  - Others:
    - Normal Tension Glaucoma
    - Toxic Maculopathy

- **Diagnosis and Discussion:**
  - **Final Diagnosis: Lupus Associated Bilateral Optic Neuropathy**
    - Optic neuropathy in lupus patients can classified as either optic neuritis or ischemic optic neuropathy. This patient presented with a painless, gradual decrease in vision over a period of time, therefore an ischemic etiology is more probable. Ischemic
optic neuropathy in lupus is usually the result of a vaso-occlusive event involving the short posterior ciliary arteries. Patients are often asymptomatic but may be found to have a subtle visual field defect before any dramatic reduction in visual acuity.

- Although this patient had previously been diagnosed with toxic maculopathy secondary to previous Plaquenil use, the integrity of the macula and OCT scans did not correspond with the typical presentation of toxic maculopathy.

**Treatment /Management and Bibliography:**

- **Treatment and response to treatment:**
  - Standard treatment includes IV and PO steroids. Dosing depends on the severity of presenting illness.
  - Improvement is usually seen within 3 weeks of initial treatment, if diagnosed early in the disease process.
  - Research has shown that IV pulse therapy of cyclophosphamide for 6 months has been effective in combination with PO steroids and/or immunosuppressants.
  - Tapering of steroids is significant and can take up to 3 years.
  - Recent studies have evaluated the occurrence of glaucomatous-like nerve damage in patients with an autoimmune disease. Immunoopathogens have been found to cause damage to the optic nerve or its vessels by attacking antigens that have cross reactivity with a primary stimulus related to the autoimmune disorder. Nerve damage is more prevalent in those never reported as having an increase in IOP.
  - This patient was prophylactically started on Travatan Z 0.004% qhs OU

- **Bibliography:**

• Conclusion:
  o This case demonstrates the power of ancillary testing and differential diagnosis in a case of lupus associated optic neuropathy.
  o Proves the value of baseline ancillary testing on patients that are at risk of ocular damage secondary to a systemic disease.
  o Shows the significance of doctors fully understanding the full scope of a disease and make an appropriate referral. In this case, obtaining an ERG, VEP and performing color vision testing would have been very beneficial in reaching a definitive diagnosis sooner.