Unilateral, sequential optic nerve edema (OD then OS) in an elderly Type 2 diabetic patient with associated pre-retinal hemorrhage: Diabetic papillopathy or incipient Anterior Ischemic Optic Neuropathy?

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Abstract: A 72 year old Caucasian male presented to clinic with unilateral diabetic papillopathy and subsequent pre-retinal hemorrhage with no proliferative retinopathy. Two years later the patient returned with unilateral diabetic papillopathy in the other eye.

I. Case History
   A. Patient demographics
      ▪ 72 year old Caucasian Male
   B. Chief complaint
      ▪ States “vision is horrible at distance and near”
   C. Ocular history
      ▪ s/p Blepharoplasty OU due to Dermatochalasis OD>OS
      ▪ DM Type II no retinopathy OU
      ▪ Choroidal Nevus OD
      ▪ Dry Eye Syndrome
      ▪ Pseudophakia OU
   D. Medical history
      ▪ Diabetes Mellitus Type II
      ▪ Hypertension
      ▪ Hyperlipidemia
      ▪ Coronary Artery Disease
      ▪ Obstructive Sleep Apnea
      ▪ Hypoplastic Anemia
      ▪ Migraine Headaches
      ▪ Gouty Arthritis
      ▪ Charcot Arthropathy
      ▪ Obesity
   E. Medications
      ▪ Allopurinol 100mg BID
      ▪ Alprazolam 0.5mg TID
      ▪ Cyanocobalamin 1000mcg QD
      ▪ Darbepoetin Alfa 200mcg/0.4mL injection every 2 weeks
      ▪ Ferrous Gluconate 325mg QD
      ▪ Furosemide 40mg BID
      ▪ Gemfibrozil 600mg BID
      ▪ Insulin nph Human 100 unit/mL Novolin 65 units BID
      ▪ Metoprolol Succinate 50mg TID
      ▪ Multivitamins
      ▪ Omeprazole 20mg QD
- Sertaline 100mg BID
- Simvastatin 40mg (1/2 tablet) QD
- Urea 20% cream (for dry skin on feet)

F. Other salient information
- BMI: 38.6
- BP (before OD ONH edema): 130/70; BP (before OS ONH edema): 124/72

II. Pertinent findings
A. Clinical
   a. First Visit/Second visit:
      1. BCVA 20/40 OU
      2. PERRL (-) APD
      3. EOMs: FROM (-) diplopia/pain
      4. HVF OD: scattered defects temporally and inferiorly; OS: scattered defects inferior nasal and superior temporal
      5. OCT Optic Disc: Nerve fiber layer thickened superior, inferior, and temporal but still within normal limits; OS: within normal limits
      6. Edematous disc with flame hemes OD
      7. 1 DX: Acute Ischemic Optic Neuropathy, due to Diabetes OD
      8. 2 DX: DM Type II Mild NPDR OU
   b. Third Visit:
      9. IVFA: OD: Primary eye transit time: 25.9 seconds (Slow); Markedly hyperfluorescent optic nerve that increases in hyperfluorescence with hypofluorescent hemes, multiple hyperfluorescent microaneurysms; NO evidence of NVD or NVE
         OS: multiple hyperfluorescent microaneurysms with few in FAZ; NO evidence of NVD or NVE
   c. Fourth Visit:
      ▪ VA (sc)- OD: 10/300 with Feinbloom PH: 10/200; OS: 20/40 PH: 20/25
      ▪ PERRL (-) APD
      ▪ D-15 Color Vision: within normal limits OD, within normal limits OS
      ▪ HVF 30-2- OD: (MD -13.79) superior temporal quadrantanopsia spilling into superior nasal field and inferior nasal step (attributable to pre-retinal heme); OS: superior arcuate due to lid
      ▪ Massive pre-retinal boat shaped hemorrhage and marked optic nerve edema with fibrotic tissue adjacent to superior nerve head OD
      ▪ 1 Dx: Large pre-retinal hemorrhage anterior to macula OD. Due to valsalva in patient with edematous optic nerve or proliferative diabetic retinopathy. RTC x 1 week for IVFA
   d. Fifth Visit:
      ▪ VA(cc)- OD: 10/300 with Feinbloom OS: 20/50
- Color/Red Free Photos- OD: swollen, congested nerve head; large pre-retinal heme with settling RBC inferiorly in boat shape, some settling superior to nerve; white fibrotic clump superior to nerve head vitreal clot
- IVFA- OD: Primary eye transit time: < 21 seconds; Markedly hyperfluorescent optic nerve with telangiectatic vessels, hypofluorescence of posterior pole due to blockage from pre-retinal heme; NO evidence of NVD or NVE; OS: multiple hyperfluorescent microaneurysms; NO evidence of NVD or NVE
- 1 Dx: Large pre-retinal hemorrhage OD likely due to bleed from chronically congested optic nerve x 4.5 months. No signs of proliferative diabetic retinopathy
- 2 Dx: DM Type II with Moderate NPDR OU
- 3 Dx: DM Papillopathy OD

e. Sixth Visit:
- VA(cc)- OD: 10/140 with Feinbloom OS: 20/25
- PERRL (-) APD
- B-Scan- OD: No mass lesion or retinal detachment; OS: normal
- A1C: 9.2

f. Seventh Visit:
- VA(sc)- OD: 20/100 OS: 20/30
- A1C: 9.5
- Resolving pre-retinal heme with chronic optic nerve head edema OD (x 7.5 months)

h. Eighth Visit:
- VA(cc)- OD: 20/40 PH: 20/30 OS: 20/25
- Resolving pre-retinal heme with chronic optic nerve head edema OD (x 10 months)

i. Ninth Visit (11 months from 1st)/Tenth Visit (17 months from 1st)/Eleventh Visit (22 months from 1st):
- BCVA- OD: 20/40 OS: 20/25
- PERRL (-) APD
- 1 Dx: Moderate NPDR OU
- 2 Dx: Fully Resolved pre-retinal heme and diabetic papillopathy OD

j. Twelfth Visit (26 months from 1st visit):
- VA(cc): OD: 20/25 OS: 20/20
- A1C: 6.6
- Edema, hyperemia of superior nerve with adjacent flame hemes OS; linear pre-retinal heme along inferior temporal arcade OS
- OCT Macular Cube: OD: no macular edema, WNL; OS: foveal elevation due to traction of vitreous (evolving PVD)
- 1 Dx: DM Type II with Moderate NPDR OU
2 Dx: Early Diabetic optic papillopathy OS with preretinal heme

k. Thirteenth Visit:
- IVFA: OS: Primary Eye transit time: 28 seconds; trace hyperfluorescence of superior ONH which increases throughout, hypofluorescence of flame hemes and pre-retinal hemes described above; hyperfluorescence of scattered microaneurysms superior/inferior temporal arcade; NO evidence of NVD or NVE; OD: hyperfluorescence of scattered microaneurysms superior/inferior temporal arcade; small area of expanding hyperfluorescence along inferior temporal arcade 2’ leaking MAs; NO evidence of NVD or NVE
- 1Dx: DM Type II with Moderate NPDR OU with early Diabetic Papillopathy OS and H/O Diabetic Papillopathy OD

l. Fourteenth Visit:
- OCT Optic Disc- OD: within normal limits, thickest 4:00-5:00 OS: thickening all quadrants, thickest 10:00-2:00 and 6:00-7:00
- 1Dx: DM Type II with Moderate NPDR OU with early Diabetic Papillopathy OS and H/O Diabetic Papillopathy OD

m. Diabetic Papillopathy OS resolved 8 months later

B. physical
- negative headache, myalgia, jaw claudication, temple pain

C. laboratory studies
- Visit #1- A1C: 8.1, SED Rate: 51 (high)- upon consultation with Rheumatology MD, due to patient’s Gout, normal CRP (2.28), normal platelets
- 5 months later- A1C 9.2, Sed Rate 60 (high), normal CRP (3.66)

D. radiology studies
- Head CT scan w/o contrast: No acute abnormality, no intracranial hemorrhage, extensive chronic ischemic microvascular disease with no significant change in an old lacunar infarct in the anterior limb of left internal capsule

E. others
- Exophthalmometry: Base 108; OD 17, OS 17

III. Differential Diagnosis
A. Primary/leading
- Diabetic Papillopathy

B. Others
- AION
- NAION
- Central Retinal Vein Occlusion
- Optic nerve compression from space occupying lesion
- Infiltrative leukemia
- Multiple Sclerosis
- Tuberculosis
- Neuroretinitis (Lyme disease, Bartonella (cat-scratch), Sarcoidosis, Syphilis, Toxoplasmosis, systemic lupus erythematosus, Wegener’s granulomatosis)
- Grave’s ophthalmopathy
- Amiodarone induced optic neuropathy
- Pseudopapilledema (i.e. Optic Nerve Head Drusen)
- Malignant Hypertensive Retinopathy
- Leber’s Optic Neuropathy
- Papillophlebitis
- Toxic Optic Neuropathy (methanol, ethambutol, ethylene glycol, and other toxins)
- Other causes (low IOP, ocular injury or blood loss, radiatation, chronic intraocular inflammation after eye surgery)

IV. Diagnosis and discussion
A. elaborate on the condition
- Diabetic Papillopathy typically presents bilaterally in young type I [insulin dependent] diabetes mellitus, irregardless of stage of diabetic retinopathy. The clinical appearance is optic disc edema that may be sectoral or total with associated circumpapillary superficial hemorrhages. Sometimes there may be surface telangiectasia on the surface of the optic nerve that can be mistaken for neovascularization. The optic disc edema may be unilateral or bilateral and may be simultaneous or sequential. Often there are no signs or symptoms associated with diabetic papillopathy. Vision loss may be mild to moderate and visual field loss varies from an enlarged blindspot to isolated scotomas.
- The pathophysiology of diabetic papillopathy is unknown, however microangiopathy has been proposed as an etiology. There is a theory that diabetic papillopathy is a mild form of nonarteritic anterior ischemic optic neuropathy (NAION) with reversible ischemia. This is due to the fact that some patient’s with diabetic papillopathy develop irreversible vision loss and optic disc pallor similar to NAION or develop NAION in the fellow eye. Diabetes is a risk factor for both entities and small optic cups are frequently seen in both diabetic papillopathy and NAION. The pathophysiology of NAION is due to acute ischemia of the posterior ciliary arteries in the optic nerve head, which is most commonly seen due to a transient fall in blood pressure.

B. expound on unique features
This case is unique due to the recurrent nature and the unilateral presentation OD first then 2 years later OS in a 72 year old type II diabetic patient.

IV. Treatment, Management
- The management is to rule out all other possible diagnoses, especially the vision/life threatening ones.
- No treatment is necessary. With diabetic papillopathy the optometrist should monitor closely with dilated fundus exams to confirm the expected resolution of the optic disc edema and fluorescein angiography to look for signs of proliferative retinopathy, such as neovascularization of the disc or neovascularization elsewhere and should educate the patient about control of blood sugar levels. Diabetic papillopathy typically resolves spontaneously within 6 months. However, it can take up to 1 year for optic disc swelling to resolve.

B. bibliography, literature review encouraged

III. Conclusion
A. clinical pearls, take away points if indicated
- Diabetic papillopathy is a diagnosis of exclusion
- It is most commonly bilateral in young type 1 diabetics
Some clinicians believe that Diabetic Papillopathy is a unique diagnosis, and others believe that it is a form of mild, reversible NAION.

You must rule out a number of other etiologies such as papilledema, space occupying lesions, and anterior ischemic optic neuropathy (arteritic and non-arteritic) before you diagnose diabetic papillopathy.