Chronic Macular Edema due to Idiopathic Juxtafoveal Telangiectasia (IJT): An Overlooked Diagnosis

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Abstract: Idiopathic Juxtafoveal Telangiectasias (IJT) is an overlooked diagnosis due to its insidious onset and similarity to other vascular retinal pathologies. This poster presents a subclinical case of IJT, diagnosis and management will be discussed.

I. Case history
   • 58 year old white male
   • Chief Complaint:
     o Fluctuation of visual acuity OS
     o Mild chronic macular edema OS with exudates and microaneurysms starting in 1994 presumed clinically significant macular edema (CSME)
   • Ocular History
     o History of Focal and Grid Laser OS in 1994 and 2004
     o History of mild Non-Proliferative Diabetic Retinopathy (NPDR) OU
     o Mixed mechanism glaucoma OU on treatment with Travoprost Z qhs OU and history of Peripheral Iridotomy (PI) OU due to narrow angles
   • Medical History
     o Hyperlipidemia
     o Hypertension
     o Diabetes Mellitus Type-1 with history of inadequate blood sugar control
     o Tobacco use
   • Medication
     o Simvastatin 20MG Tab QD
     o HCTZ 25/Lisinopril 20MG Tab QD
II. Pertinent findings

- Clinical:
  - Best Corrected Visual Acuity 20/20 OD, 20/20- OS
  - Pupils within normal limits
  - Anterior Segment:
    1. Patent superior temporal PI OD/OS
    2. IOP: 14 mmHg OD and 15 mmHg OS
    3. Anterior chamber is deep and quiet
  - Fundus Exam:
    1. Vitreous:
       a. Syneresis OU
       b. No blood, cells, or pigment present
    2. Optic nerves:
       a. OD: 0.60/0.45 OD pink and healthy
       b. OS: 0.45/0.45 OS pink and healthy
    3. Vasculature: 2/3 AV ratio, no venous beading, no IRMA, no hypertensive changes
    4. Macula
       a. OD:
          i. 4-5 juxtafoveal dot hemorrhages, no CSME
       b. OS:
          i. 4-5 juxtafoveal dot hemorrhages with exudates and retinal thickening within 500 microns of the fovea
          ii. Multiple, large micro-aneurysms at arteriolar endpoints, vascular out pocketing at the macula, and telangiectatic vasculature limited to the juxtafoveal region
    5. Periphery: Few scattered dot hemorrhages OD and OS

- Laboratory Studies
  - Hemoglobin A1C 8.4%

- Imaging
  - Fluorescein Angiography (FA):
    1. OD: Few hyperfluorescent microaneurisms near the macula no CSME or leakage noted
    2. OS: Hyperfluorescent microaneurisms and spotted leakage extending into the foveal avascular zone corresponding to the telangiectatic vessels, no areas of ischemia no note of true CSME leakage
  - OCT
    1. OD: regular foveal contour, normal foveal depression
    2. OS: slightly flattened retinal contour, intra-retinal cystic edema centrally

III. Differential diagnoses
• Primary: IJT
• Others:
  o Mild NPDR with CSME
  o Old Branch Retinal Vein Occlusion (BRVO)
  o Prostaglandin induced cystoid macular edema (CME)
  o Choroidal neovascular membrane (CNVM)

IV. Diagnosis and discussion
• Chronic macular edema and exudates indicate leakage of the juxtafoveal vasculature
• The incidence of diabetes in patients with IJT is reported to be similar to the incidence in the general public with underlying ischemia being common to both
• Most likely not CSME due to
  o Minimal leakage concentrated to the macula during the FA
  o Leakage is spotted and corresponding to outpocketing of vessels seen juxtafoveally
  o FA is crucial in the diagnosis as leakage was confined to areas around the telangiectatic vessels
• No sclerosed vessels or areas of retinal ischemia ruling out BRVO

V. Treatment and management
• No treatment is indicated at this time and patient will be followed closely for any progression
• Patient education that the condition is an insidious, progressive disease that can lead to significant visual complications in the future
• Patient given a home Amsler Grid to self-monitor for central vision changes due to macular edema
• Focal and grid laser photocoagulation are mainstay treatments for CSME but are only indicated in select cases of IJT
• Standardized treatment of early to advanced IJT is un-established
• Multiple small studies have attempted a variety of therapeutic modalities (laser photocoagulation, steroid injections, anti VEGF injections, etc), each having mixed or ineffective results
• Currently a multicentre, large scaled study is underway regarding best practices in the treatment of IJT