I. Case History

□ Demographics: 43-year-old white male

□ Chief complaint: Notices floaters (possibly only in the left eye) for about 3-4 months. (-) flashes. Distance vision is fine unaided. Uses readers for near work.

□ Ocular history: unremarkable; (-) injury, (-) surgery

□ Medical history:
  h/o smoking
  h/o cocaine use
  nodule of the lung; CT x 2010
  sleep apnea; (+) CPAP
  GERD
  Bipolar, anxiety, depression
  MRSA
  hyperlipidemia

□ Medications:
  Colchicine
  Allopurinol
  Pantoprazole
  Hydrocodone

II. Pertinent Findings

⇒ Clinical:
  o Best corrected acuity: 20/20 OD, OS
  o Pupils: d/c; (-)APD
  o Preliminary testing: Color vision (Ishihara): 14/14 correct OD, OS
    Blood pressure: 126/78 right arm, sitting
  o Slit lamp exam: unremarkable OU
  o Tonometry: 12mmHg OD, OS
Dilated fundus exam:

- OD: unremarkable
- OS:
  - Cup/disc ratio: 0.15h/0.25v, 2-3+ hyperemia 360 degrees, sectoral disc edema of superior temporal rim and adjacent nerve fiber layer from 12 to 1:30 o’clock, (-) hemes, (-) exudates, (-) telangiectasia, shunt, or feeder vessels
  - Macula: normal
  - Vessels: normal, (-) sheathing
  - Periphery: normal

Laboratory studies: ANA, CRP, RPR, FTA-ABS, ACE, serum lysozyme, CBC; all normal

Imaging studies:
- MRI brain/orbit with contrast: no mass, no hemorrhage, no demyelinating plaques.
- Longitudinal studies over 5 years with Heidelberg OCT and accompanying fundus photography. Superior optic nerve swelling OS that resolves over time (show tumor regression)
- Visual fields: scattered superior temporal defects OS with poor reliability and repeatability, no correlation to optic nerve changes
- Supporting FA: focal enhancement area overlying the superior temporal portion of the left disc consistent with capillary hemangioma

III. Differential diagnosis

- Leading: NAION, optic disc granuloma, optic disc glioma

- Others: Giant cell arteritis, optic neuritis, compressive optic neuropathy, inflammatory optic neuropathy, infectious optic neuropathy, traumatic optic neuropathy, toxic optic neuropathy, infiltrative optic neuropathy, optic nerve head drusen, papilledema
IV. Diagnosis & Discussion

Juxtapapillary retinal capillary hemangiomas (JRCHs) are benign vascular tumors that are classified as endophytic and arising from the inner retinal layers or exophytic which arise from the outer retinal layers. JRCHs most often present in the endophytic form and are easily identified during dilation. Typical features of endophytic JRCHs include presence of dilated tortuous feeding artery with a draining vein and these are commonly associated with von Hippel Lindau disease (VHL)\(^3\). The diagnostic challenge arises when the tumors are exophytic and the classical tumor appearance is not observed. Exophytic JRCHs are often misdiagnosed as papilledema, papillitis or granulomatous disease\(^4\). Recent studies show that OCT, fluorescein angiogram, MRI and a thorough family history can help with the diagnosis of exophytic JRCHs. The MRI of the brain and spinal cord confirms the presence or absence of other central nervous system tumors which are common with VHL. An OCT reveals a round lesion with shadowing but unlike other ocular tumors, with a clear delineation of the normal overlying or underlying retina depending on the location of the tumor\(^3\). The FA shows early filling of the retinal vessels, mid phase filling of the hemangioma and late phase excessive leakage from the tumor\(^4\). Any family history of VHL should be elicited due to a high correlation with retinal hemangiomas.

V. Treatment & Management

Treatment of this JRCH tumor was not indicated since asymptomatic in nature with 20/20 visual acuity OU and lack of other complications. Potential complications can include subretinal fluid accumulation, exudative retinal detachment, macular edema, or epiretinal membrane formation that can lead to a tractional retinal detachment\(^1\). Treatments in symptomatic patients with exophytic lesions are challenging due to the juxtapapillary location of the hemangioma. Treatment outcomes are often poor for exophytic tumors due to problems of treatment delivery deep into the outer retina and can lead to scotomas post treatment without improvement in the condition\(^1\). Some treatment options include anti-VEGF, PDT and intravitreal surgery\(^1\). Saitta et al presents an algorithm approach for JRCH treatment that can be easily followed based on the location of the tumor and presence of complications.
VI: Conclusion

JRCHs are benign tumors that can be associated with VHL or occur sporadically. They may cause complications that can lead to poor visual outcome. Treatment should be considered since most are progressive\(^1\). Observation is a viable option when the patient is asymptomatic, especially when the location of the tumor is juxtapapillary and treatment options are limited and often unsuccessful. Regression of these tumors is uncommon. Fortunately, the JRCH regressed and no treatment was necessary in our patient. Due to the longstanding and stable condition of the JRCH, the patient is being monitored with annual dilation, OCT and fundus photography. Although solitary JRCHs are difficult to diagnose, it is important for clinicians to consider this condition when presented with disc edema of unknown etiology and further investigate the patient case.

VII. Bibliography
