The use of OCT angiography in two cases of confirmed idiopathic intracranial hypertension

Samantha J. Rao, O.D.
Primary Care/Cornea and Contact Lens Resident
Nova Southeastern College of Optometry

ABSTRACT: This case series investigates the features of optic disc edema as seen on OCT angiography. Identification of these distinct features can be valuable in differentiating the various optic neuropathies that present with optic disc swelling.

I. CASE HISTORIES
   a. Patient 1
      i. 20 year old African-American female
      ii. Patient presented to the clinic complaining of blurry and occasional double vision, severe headaches, tinnitus, numbness and tingling in her upper and lower extremities. Her symptoms began shortly after starting Levora oral contraceptive three months prior as prescribed by her gynecologist.
      iii. Her ocular history was unremarkable. Her medical history was significant for polycystic ovarian syndrome. Not currently taking any other medications.
   b. Patient 2
      i. 26-year-old African-American female
      ii. Patient presented complaining of blurry vision OD>OS and darkened vision upon awakening in the morning which gradually lightened in patches as she stood up. This had occurred every morning for the past two weeks. She also described a foggy veil over right eye for the past two weeks, and complained of ringing in her right ear. She was not experiencing headaches.
      iii. Her ocular and medical histories were unremarkable. No current medications.

II. PERTINENT FINDINGS
   c. Patient 1
      i. Best corrected distance VA’s:
         1. OD: 20/400+1
         2. OS: 20/400+1
      ii. Normal confrontation visual fields, EOMs, pupils; no RAPD; clear media
      iii. IOP OD: 18 mmHg, OS: 17 mmHg
      iv. Blood pressure: 125/80 mmHg
      v. Dilated fundus examination
         1. OD: Swollen, hyperemic optic discs with elevated and blurred margins 360 and scattered papillary splinter hemorrhages concentrated nasally; Paton’s folds extending temporally toward macula. Tortuous vessels surrounding optic disc. Macula flat and intact with foveal light reflex.
         2. OS: Swollen, hyperemic optic discs with elevated and blurred margins 360; two small cotton wool spots nasally; Paton’s folds. Tortuous vessels...
surrounding optic disc. Macula flat and intact with foveal light reflex.

vi. Imaging
1. SD-OCT: Profound increase in RNFL thickness in all quadrants OU. Normal foveal contour; no macular edema.
2. Visual field (FASTPAC Size V): Mildly enlarged blind spot OU.
3. OCT-Angiography (Zeiss Angioplex):
   a. Optic Nerve (6x6): Increased visibility of the peripapillary capillary network, dilated and tortuous peripapillary and prelaminar vasculature, and a dense microvascular network visible over the optic nerve head.
   b. Macula (6x6): Normal foveal avascular zone and surrounding vasculature OU.

b. Patient 2
i. Best corrected distance VA’s:
   1. OD: 20/20
   2. OS: 20/20
ii. Normal confrontation visual fields, EOMs, pupils; no RAPD; clear media
iii. IOP OD: 13 mmHg, OS: 14 mmHg
iv. Blood pressure: 117/70
v. Dilated fundus examination
   1. OD: Mild diffuse optic disc hyperemia with elevation and blurring of disc margins; Paton’s folds, no hemorrhages. Tortuous vessels surrounding optic disc. Macula flat and intact with foveal light reflex.
   2. OS: Mild diffuse optic disc hyperemia with elevation and blurring of disc margins; Paton’s folds, no hemorrhages. Tortuous vessels surrounding optic disc. Macula flat and intact with foveal light reflex.

vi. Imaging
1. OCT-Angiography (Optovue AngioVue)
   a. Optic Nerve (6x6): Increased visibility of the peripapillary capillary network, dilated and tortuous peripapillary and prelaminar vasculature, and a dense microvascular network visible over the optic nerve head.
   b. Macula (6x6): Normal foveal avascular zone and surrounding vasculature OU.

III. DIFFERENTIAL DIAGNOSES
   e. Idiopathic intracranial hypertension
   b. Venous sinus thrombosis
   c. Cerebral neoplasm, aneurysm, hematoma or hemorrhage
   d. Compressive optic neuropathy (e.g. aneurysm, neoplasm)
   e. Infiltrative optic neuropathy (e.g. neoplasm, leukemia, sarcoidosis, tuberculosis)
   f. Infectious papillitis, meningitis or encephalitis (e.g. Lyme disease, syphilis)
   g. Optic neuritis (e.g. multiple sclerosis or other autoimmune disease)
   h. Non-arteritic anterior ischemic optic neuropathy (NAION)
IV. DIAGNOSIS AND DISCUSSION

Both patients were sent to the emergency room that same day for an MRI of brain and orbits, MRA, MRV, lumbar puncture, ESR, CRP and CBC with differential. In both cases, all imaging and serological testing were unremarkable, but both patients had elevated opening pressures on lumbar puncture. The first patient’s opening pressure was 46 cmHg, and the second patient’s opening pressure was 36 cmHg. Both patients were diagnosed with idiopathic intracranial hypertension (IIH).

Although there are numerous studies reporting the use of OCT angiography (OCT-A) for evaluation of the retinal and choroidal structures, few studies have evaluated optic nerve head vasculature. In cases of acute idiopathic intracranial hypertension, OCT-A will likely reveal increased visibility of the peripapillary capillary network, dilated and tortuous peripapillary and prelaminar vasculature, and a dense hypervisible microvascular network over the optic nerve head\(^1\). The scan may show a dark ring around the optic nerve which may be misinterpreted as a reduction in perfusion in that area due to swelling, but in fact the dark ring is merely an artifact created by the fact that the swollen optic nerve is in a much higher plane than the surrounding retina. As disc edema begins to resolve, the increased visibility and dilation of the peripapillary and prelaminar vasculature should begin to return to normal. Optimal visualization is achieved via simultaneous viewing of the RNFL on SD-OCT and OCT-A; patients with disc edema due to IIH will demonstrate profuse NFL thickening on SD-OCT.

Miscellaneous optic neuropathies including NAION may present with segmental involvement which will manifest as sectoral RNFL elevation on the SD-OCT in the acute phases. OCT-A will highlight areas of active edema as an increase in visibility of the radial peripapillary vessels with coinciding dilation and tortuosity of the adjacent vasculature. Over time, the nerve atrophies and the OCT-A will demonstrate a reduction in visibility of the peripapillary microvessels\(^2\).

V. TREATMENT AND MANAGEMENT

Both patients were started on Acetazolamide 250mg BID by mouth and followed up with neuro-ophthalmology. At a 4-day follow-up, Patient 1’s vision had improved to 20/200 OD and OS and her neurological symptoms and headaches were beginning to resolve. At a 1-month follow-up, Patient 2’s vision continued to be 20/20 OD/OS and her neurological symptoms had mostly dissipated, but she was still experiencing some persistent ringing in her right ear.

VI. CONCLUSIONS

OCT-A in conjunction with SD-OCT provides an adjunct means of visualizing the microvascular peripapillary network and can provide diagnostic information alongside information about the chronicity of the neuropathy. Herein we present the OCT-A findings seen in two cases of confirmed IIH with varying degrees of involvement and alterations in the peripapillary perfusion.

VII. WORKS CITED