Macular Optical Coherence Tomography Findings in Patients with Syphilitic Optic Neuropathy – A Case Series and Systematic Review

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Introduction: Syphilis is a sexually or congenitally acquired infectious disease that can affect multiple organs systems, including the eye. When left undiagnosed and untreated, it can lead to significant morbidity and mortality. Syphilitic optic neuropathy can be difficult to diagnose as it can mimic many other non-syphilitic causes of optic-nerve involvement, leading to delay in treatment. Diagnosing ocular syphilis may be facilitated by assessing for specific outer retina abnormalities on macular ocular coherence tomography (OCT).

Methods: This was a case series and case-based systematic review. For the case series, a retrospective chart review was conducted of all patients who presented to a tertiary university-affiliated neuro-ophthalmology practice over 6 months with undifferentiated optic neuropathy and were eventually diagnosed with syphilitic optic neuropathy. For the systematic review, OVID MEDLINE, EMBASE and COCHRANE CENTRAL databases were searched to identify all cases of syphilitic optic neuropathy with macular OCT. The primary research outcome was the prevalence of cases with outer retinal abnormalities on macular OCT.

Results: Four cases were identified that were eligible for inclusion. The ages ranged from 27 to 62 years old, and two of the patients were female. On examination, vision ranged from Snellen 20/50 to hand motion; all patients had optic neuropathy and macular OCT revealed chorioretinitis characterized by retinal pigment epithelium excrescences. The patients subsequently underwent uveitis work-up and were diagnosed with syphilis. They were treated with intravenous penicillin and showed improvement in outer retina appearance on follow-up. The systematic review consisted of 24 cases and 35 eyes with syphilitic optic neuropathy and reported macular OCT findings. Eighty-three percent (20/24) were males, and the mean age was 47.7 (SD: 49.2). The mean visual acuity at presentation was Snellen 20/57. On fundoscopy, 25.7% (9/35) of eyes had vitritis, while 22.8% (8/35) had placoid chorioretinal lesions. On OCT, 45.7% (16/35) of eyes had abnormal outer retina findings, most commonly disruption of the ellipsoid zone and/or retinal pigment epithelium excrescences. All patients were treated with penicillin or ceftriaxone, and final mean visual acuity was Snellen 20/29.

Conclusions: Two patients identified in the case series and nearly half of patients in the literature with syphilitic optic neuropathy had concurrent specific outer retina abnormalities (disruption of ellipsoid zone and/or placoid chorioretinitis in the form of RPE excrescences) seen on macular OCT. Thus, we recommend clinicians obtain macular OCT for all patients presenting with undifferentiated optic neuropathy to facilitate earlier diagnosis of syphilis.