Optical Coherence Tomography in Neuro-ophthalmology

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KEYWORDS

• Optical coherence tomography • Optic neuritis • Neuromyelitis optica
• Multiple sclerosis • Optic disc drusen • Papilledema
• Idiopathic intracranial hypertension • Pituitary tumors

KEY POINTS

- The diagnosis of many central nervous system (CNS) disorders can be facilitated through a detailed ophthalmic examination and adjunctive optical coherence tomography (OCT).
- OCT represents a surrogate marker of neuroaxonal integrity in the afferent visual pathway. Neurodegenerative components of CNS diseases can be captured with this ocular imaging technology.
- OCT should be viewed as a complement to, but not a replacement for, function visual outcomes in patients with CNS disorders.
- OCT has proved a useful tool in monitoring demyelinating disorders of the CNS, distinguishing causes of optic nerve elevation, and diagnosing pituitary tumors.

INTRODUCTION

Cardinal features of many CNS disorders can be identified through a detailed fundus examination. In the setting of raised intracranial pressure, the optic nerve may appear edematous (ie, papilledema), the severity of which has been classified according to grading scheme (eg, the Frisen scale).[1] In cases of chronic optic neuropathy secondary to compressive, ischemic, inflammatory, infiltrative, and infectious etiologies, the optic nerve often appears pale and atrophic. Retinal nerve fiber layer (RNFL) defects may manifest as a consequence of an optic nerve injury. Yet, these fundus features can be difficult to identify, even by experienced observers. In the modern era, OCT

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complements conventional ophthalmoscopy techniques, by providing a quantitative means of capturing acute and chronic effects of optic nerve injury. The RNFL is comprised, in part, by retinal ganglion cell axons and lacks myelin. Therefore, changes in RNFL thickness have been interpreted to reflect axonal injury.[2] Within the macular region, there is a high proportion of retinal ganglion cells. Loss of macular thickness and thinning of the ganglion layer represent neuronal damage in afferent visual pathway. In recent years, OCT has emerged as a structural marker, which may facilitate understanding of mechanisms of neurodegeneration that contribute to neurologic disability in a variety of CNS diseases.

MONITORING DEMYELINATING DISORDERS OF THE CENTRAL NERVOUS SYSTEM

Acute Optic Neuritis

Optic neuritis may occur as a sporadic event or in the context of multiple sclerosis (MS). For 1 in 5 MS patients, optic neuritis represents the first clinical manifestation of their disease.[2] The Optic Neuritis Treatment Trial demonstrated that most optic neuritis patients are young (mean age: 32 years), white (85%), and women (77%) who report pain at the time of symptom onset (92%).[2–4] Vision loss tends to progress over hours to days and may range from mild (Snellen visual acuity equivalent of 20/20) to severe (no light perception) initially.[2] In patients with unilateral optic nerve involvement, a relative afferent pupil defect (RAPD) can be detected in the affected eye. In the context of bilateral optic neuritis, the RAPD localizes to the more severely affected eye but an RAPD may be absent in bilateral and symmetric disease. Patients frequently note color desaturation, referred to as dyschromatopsia, which can be disproportionate to their high-contrast visual acuity deficit.[2] In cases of retrobulbar optic neuritis, the fundus examination may appear normal, whereas patients with anterior optic neuritis (papillitis) demonstrate mild to moderate optic disc swelling.[2] Visual recovery after optic neuritis typically occurs within weeks, and the overall prognosis is favorable, with more than 90% of patients achieving a visual acuity of 20/40 after 1 year.[2–4] Despite regaining normal visual function as measured by standard ophthalmic testing, many optic neuritis patients report persistent problems with heat-induced vision loss (Uhthoff phenomenon), altered motion perception, and decreased spatial vision at low-contrast levels. There is, therefore, a need for more sensitive measures of structural injury and functional impairment in this patient population.

In the acute phase, OCT-measured peripapillary RNFL measurements are often elevated in the optic neuritis eye, presumably due to axoplasmic flow stasis.[2,5] This initial spike in RNFL thickness makes it difficult to identify the earliest signs of retrograde axonal degeneration from the retrobulbar site of optic nerve inflammation and hampers attempts to precisely tract axonal injury. In contrast, OCT-measured macular volume and ganglion layer measures are generally comparable between affected and unaffected eyes of patients at the time of symptom onset, declining for 6 to 12 months thereafter.[5] Retinal segmentation techniques have shown that in the initial months that follow optic neuritis, the percentage decrease in ganglion layer thickness correlates with increased outer nuclear layer and photoreceptor layer thicknesses, albeit these outer layer measurements subsequently decline between months 4 and 12 postevent.[6] Postacute functional outcomes (high-contrast and low-contrast letter acuity, color vision, and visual field sensitivity measures) after optic neuritis correlate with the eventual of extent of OCT-measured RNFL, ganglion layer, and macular volume loss detected 6 to 12 months after the event.[2]

Recurrent optic neuritis has been associated with worse OCT measures of neuroaxonal integrity in the afferent visual pathway. Yeh and colleagues[7] demonstrated a