

CANADIAN NEURO-OPHTHALMOLOGY SOCIETY

THURSDAY 12 JUNE

Paper #A-00048

Tracking retinal nerve fiber layer changes after optic neuritis

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Purpose: Optic neuritis (ON) causes retinal nerve fiber layer (RNFL) damage, which can be quantified with optical coherence tomography (OCT). We used OCT to track RNFL changes after ON, and then determine whether a time-dependent relationship exists between RNFL thickness and visual function.

Methods: In this prospective case series, 78 patients with ON as a clinically isolated syndrome (CIS) underwent serial OCT and visual testing over a mean period of 28 months. The main outcome measures were comparisons between RNFL values in clinically affected and non-affected eyes; and correlations between RNFL thickness and visual function over time.

Results: Significant RNFL thinning manifested in clinically affected eyes less than 3-months after ON, and persisted for greater than 24 months. The earliest detectable RNFL thinning occurred in the temporal region. Most thinning manifested 3-6 months after ON, after which RNFL values stabilized. Nasal RNFL values showed the least consistent differences between clinically affected and unaffected eyes over time. Temporal values were significantly thinner in affected eyes up to 24 months after ON, after which comparisons between clinically affected and unaffected eyes were less robust. Regression analyses demonstrated a threshold of RNFL thickness (75 μ m), which predicted visual field recovery up to 18 months after ON. The relationship between RNFL thickness and visual field function was most robust at 7-12 months ($p = 0.0129$). We did not identify a threshold of RNFL thickness which predicted visual acuity recovery, but the relationship between reduced RNFL values and diminished log MAR acuity was significant 6 or more months after ON ($p = 0.0001$).

Conclusions: RNFL thinning progresses for up to 6 months after ON. A follow up period of 12-months represents an optimal interval to track RNFL changes, and to correlate RNFL values with tests of visual recovery after ON.

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Paper #A-00049

Ophthalmologic features of pediatric pseudotumor cerebri

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Purpose: Although visual loss in substantial numbers of adult patients with pseudotumor cerebri is well recognized, in children the disease is considered to spare the visual system. We evaluated the ophthalmologic features and natural history of children with pseudotumor cerebri with long term follow-up.

Methods: Over a hundred cases of pseudotumour cerebri were examined with a retrospective chart review at the Hospital for Sick Children from the years 1975 to the present. The clinical variants of this syndrome, natural history of its evolution, ocular findings and managements were studied.

Results: Only a few cases showed significant visual loss. A significant number presented with less-severe but prominent abnormalities of visual acuity and/or visual fields at some point during the disease process which resolved with treatment. Some cases showed eventual optic atrophy but retained clinically normal visual parameters. Sixth nerve palsies were the most common oculomotor sign. One case developed an OTR and INO which cleared on medical management. One case which presented with advanced field loss and optic atrophy developed progressive visual loss despite prolonged and aggressive medical therapy. More patients with dural sinus thrombosis had serious visual loss than did those with other associated diseases or idiopathic pseudotumor cerebri. No ON sheath fenestrations were done. Neurosurgical LP shunt procedures were done in a few cases. Medical management treated the majority of cases successfully.

Conclusions: Ocular signs of pseudotumor cerebri are numerous, with sixth nerve palsies being most common. The visual prognosis for most patients with pseudotumor cerebri is good with treatment. Optic nerve appearance and clinical identification of RNF loss was more useful in management decision making than visual field assessments, which are less reliable in the younger age group. Ischemic changes and evidence of RNFL atrophy secondary to papilledema served as signs for more aggressive treatment.

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Autoimmune lymphocytic infundibulo-neurohypophysitis: an unusual cause of optic neuritis in a child

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Purpose: To report a rare case of a child with recurrent optic neuritis associated with autoimmune LIN.

Methods: A 13-year-old boy with recurrent optic neuritis was referred for ophthalmic evaluation. The child was subjected to detailed ocular, neurological and neuroradiologic evaluations.

Results: The patient initially presented with headache, polyuria and polydipsia which was confirmed to be due to diabetes insipidus. A few months later he complained of right visual impairment. Clinical, electrophysiological, and radiological examinations confirmed the diagnosis of retrobulbar optic neuritis. Over the next one year, he experienced three similar episodes of visual loss. Each time he was treated successfully with intravenous followed by oral corticosteroids. Magnetic resonance imaging (MRI) at the time of initial presentation and repeated 3 times over the next two years, displayed a progressively increasing thickening of the pituitary stalk. The tissue specimen resected at transnasal surgery showed diffuse meningeal and pituitary lymphocytic infiltration consistent with diagnosis of LIN. Subsequently the patient developed hypothyroidism and hypogonadism. He was commenced on azathioprine as a steroid sparing agent and hormonal supplementation with DDAVP, thyroxine and testosterone.

Conclusions: LIN is a rare condition clinically associated with headaches, visual symptoms, and signs of hypopituitarism. The radiological appearance of a sellar mass lesion mimics a pituitary tumor. Histopathological examination shows chronic inflammation with lymphocytic infiltration of the pituitary and surrounding meninges. Presence of diabetes insipidus, as in our patient suggests involvement of the infundibulum and neurohypophysis. Recurrent optic neuritis has been described in patients with this condition, but ours is the first report in a child. This condition must be considered in the differential diagnosis in patients with optic neuritis associated with symptoms and signs of pituitary insufficiency.