ABSTRACTS

Neuro-Ophthalmic Literature Review

Carmen Chan, Panitha Jindahra, Evan Price, John H. Pula, Michael Vaphiades,
An-Guor Wang

STROKE AND VISION: A COMPREHENSIVE REVIEW

Lawlor M, Perry R, Hunt BJ, Plant GT. Strokes and vision: The management of ischemic arterial disease affecting the retina and occipital lobe. *Surv Ophthalmol* 2015 Jul-Aug;60(4):296–309.

This comprehensive review written by neuro-ophthalmologists and neurologists summarizes the management of strokes which affect vision. In addition to the general management of ischaemic strokes, it also focuses on transient monocular visual loss, central/branch retinal artery occlusion, transient binocular visual loss, and posterior circulation strokes. For each condition, the presentation, diagnosis, management (acute to long-term), and prognosis are discussed. This is a very useful review for general and neuro-ophthalmologists.

Carmen Chan

ACUTE OPTIC NEURITIS TREATMENT REVIEW

Bennett JL, Nickerson M, Costello F, Sergott RC, Calkwood JC, Galetta SL, Balcer LJ, Markowitz CE, Vartanian T, Morrow M, Moster ML, Taylor AW, Pace TWW, Frohman T, Frohman EM. Re-evaluating the treatment of acute optic neuritis. *J Neurol Neurosurg Psychiatry* 2015;86:799–808.

Current and future approaches of acute optic neuritis were reviewed in this article. For early treatment, corticosteroids are still the mainstay. However, they do not prevent optic nerve atrophy. Plasma exchange, IVIg, ACTH, and erythropoietin are discussed in detail. By using retinal nerve fiber layer thickness as a surrogate measurement, some small trials show that memantine, dilantin, or amiloride might have a role in neuroprotection. Anti-LINGO1 promotes CNS remyelination in multiple sclerosis and may improve visual outcome in acute optic neuritis. Technologies in optic nerve evaluation such as MRI, electrophysiology, OCT, and SLP are included in the review.

Panitha Jindahra

ISCHEMIC OPTIC NEUROPATHY REVIEW

Biousse V and Newman NJ. Ischemic optic neuropathies. *N Engl J Med* 2015;372:2428–36

The review contains a discussion of the clinical presentation, pathophysiology, and treatment of ischemic optic neuropathies, including a review of the blood supply to the optic nerve with helpful diagrams.

Panitha Jindahra

EXPANDING THE ARMAMENTARIUM FOR TREATMENT OF BINOCULAR DIPLOPIA

Robert, MP, Bonci F, Pandit A, Ferguson V, Nachev P. The scotogenic contact lens: A novel device for treating binocular diplopia. *Br J Ophthalmol* 2015;99:1022–1024.

Soon, there may be a new tool available to the clinician struggling with the dilemma of symptomatic improvement of binocular diplopia in patients who are unable or unwilling to pursue prismatic correction, surgical correction, or total occlusion of one eye. The authors describe a “scotogenic” soft contact lens that is designed to induce a relative central scotoma in one eye by imparting a varying level of degradation of the perceived image in a pattern that inversely mirrors the physiologic variation of spatial acuity across the monocular visual field. It is argued that this method of relative degradation is superior to the total occlusion that patching provides in many ways, including the fact that peripheral vision in the eye wearing the scotogenic lens is conserved while not allowing for diplopia. In a comparison between these two methods of symptom control, both patients with binocular diplopia and normal subjects displayed statistically equal symptom control, as well as better aesthetics in the scotogenic lens group. In normal subjects, the patch and the lens were seen as equally comfortable, but the lens was judged to be more comfortable than the patch in the diplopia patients. On automated Estermann perimetry (performed only on the normal participants), the occlusive patch produced a mean of 14.92 missed locations, as compared to 0.92 missed locations in the scotogenic lens group ($p < 0.001$). The authors also note a few ways the device may be tailored to the specific patient at hand or altered to produce the same effect thorough an alternate method.

Evan Price

UPDATE ON LEBER'S OPTIC NEUROPATHY

Meyerson C, Van Stavern G, McClelland C. Leber hereditary optic neuropathy: Current perspectives. *Clin Ophthalmol* 2015 Jun 26;9:1165–76.

The authors provide a comprehensive and up-to-date review of Leber's optic neuropathy, including commentary on epidemiology, clinical features, and prognosis. Cellular aspects which are considered include mitochondrial function and oxidative phosphorylation, mitochondrial genetics, including penetrance, and optic nerve vulnerability. Therapies are also reviewed, including symptomatic and supportive treatments, nutritional supplements, brimonidine, ubiquinone analogues, and pharmacologic activators of mitochondrial biogenesis. Finally, gene therapy and stem cells receive a discussion, although the reviewers admit that many questions on treatment remain, and there are no established or curative medications at this time.

John Pula

THE ENIGMA OF PSEUDOPHAKIC DYSPHOTOPSIA

Michelson MA, Holladay JT. The intersection of optics and neuro-ophthalmology: The enigma of pseudophakic dysphotopsia. *J Neuroophthalmol* 2015;35(2):109–11.

Disturbing glittering sensations of light following cataract surgery may actually originate from the intraocular lens. In this editorial by Michelson and Holladay, this very important topic of positive and negative dysphotopsias, as well as multifocal intraocular lens intolerance, is reviewed.

Michael Vaphiades

OPHTHALMOPARESIS AND CRANIAL NERVE SCHWANNOMAS

Shin RK, Mejico LJ, Kawasaki A, Purvin VA, Moster ML, Younge BR, Bogen D. Transient ocular motor nerve palsies associated with presumed cranial nerve schwannomas. *J Neuroophthalmol* 2015;35(2):139–43.

The authors present a case series of ten cases of presumed cranial nerve schwannoma that presented with transient or recurring ocular motor nerve deficits. Since schwannomas typically present with gradually

progressive symptoms, this topic is noteworthy. In this case series, schwannomas of the third, fourth, and fifth nerves resulted in fluctuating cranial nerve deficits with persistent nodular cranial nerve enhancement on sequential cranial MRI scans. Several episodes of transient oculomotor deficits were associated with headache mimicking ophthalmoplegic migraine. The authors propose that some cases of ophthalmoplegic migraine are really this reported entity of schwannoma of the oculomotor nerve. The neuroimaging and clinical course are similar in both entities. The authors concluded that cranial nerve schwannomas may result in relapsing and remitting cranial nerve symptoms for which the etiology is unknown.

Michael Vaphiades

IPILIMUMAB OPTIC NEUROPATHY

Yeh OL, Francis CE. Ipilimumab-associated bilateral optic neuropathy. *J Neuroophthalmol* 2015;35(2):144–7.

The authors report a patient who developed bilateral optic neuropathy with disc edema while taking ipilimumab, a novel monoclonal antibody shown to improve survival in metastatic melanoma. Previous case reports have documented its association with drug-induced uveitis.

Michael Vaphiades

TOPIRAMATE-INDUCED PALINOPSIA

Yun SH, Lavin PJ, Schatz MP, Lesser RL. Topiramate-induced palinopsia: A case series and review of the literature. *J Neuroophthalmol* 2015;35(2):148–51.

The authors report a case series of 9 patients and review of the literature of palinopsia as a possible side effect of topiramate. Palinopsia is the persistence of or recurrence of visual images after the stimulus has been removed. Some of these patients experienced a “trailing” effect which the authors included in the definition of palinopsia. The visual effect resolved in most of the patients after stopping or decreasing the dose of the drug. The lowest dose of topiramate causing palinopsia was 25 mg twice a day. The authors concluded that topiramate-induced palinopsia may be under-diagnosed unless physicians inquire about palinopsia in these patients.

Michael Vaphiades

BLIND WITH OVARIAN CANCER

Mbekeani JN, Haseeb MQ, Tulbah AM, Hamed SH, Al Hazzaa SA, Dogar MA. Signet ring cell adenocarcinoma and bilateral leptomeningeal involvement of optic nerve sheaths. *J Neuroophthalmol* 2015;35(2):162–4.

The authors report a 41-year-old woman who presented with bilateral no light perception vision and bilateral optic disc edema and abdominal distention. An ovarian mass was detected and biopsy revealed signet ring cells. Contrasted cranial MRI showed widespread central nervous system and cranial nerve enhancement (carcinomatosis). Signet ring cell adenocarcinoma has a propensity for leptomeningeal carcinomatosis, and although bilateral optic nerve involvement is rare, this case illustrates its occult and devastating nature.

Michael Vaphiades

RETINAL FINDINGS IN DIC

Ginat DT, Christoforidis GA. Retinal and intracranial haemorrhage in acute leukemic disseminated intravascular coagulation. *Neurology* 2015;85:107–8.

The authors present very striking CT and MRI images of numerous multifocal cortical and subcortical intraparenchymal hematomas as well as bilateral retinal haemorrhages seen on the CT and MRI susceptibility weighted images.

DIAGNOSTIC CRITERIA FOR NMO

Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, de Seze J, Fujihara K, Greenberg B, Jacob A, Jarius S, Lana-Peixoto M, Levy M, Simon JH, Tenenbaum S, Traboulsee AL, Waters P, Wellik KE, Weinschenker BG. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology* 2015;85:177–89.

Neuromyelitis optica (NMO) is an inflammatory CNS syndrome distinct from multiple sclerosis (MS) that is associated with serum aquaporin-4 immunoglobulin G antibodies (AQP4-IgG). The authors describe how prior NMO diagnostic criteria required optic nerve and spinal cord involvement. However, more restricted or more extensive CNS involvement may occur. A new NMO nomenclature defines the term NMO spectrum disorders (NMOSD) which includes AQP4-IgG, clinical syndromes and MRI findings related to optic nerve, spinal cord, area postrema, other brainstem, diencephalic, or cerebral presentations. More stringent clinical criteria, with additional neuroimaging findings, are required for diagnosis of NMOSD without AQP4-IgG or when serologic testing is unavailable.

Michael Vaphiades

CHIASMAL GERMINOMA

Chaudhry NS, Ahmad FU, Whittington E, Schatz N, Morcos JJ. Primary intrinsic chiasmal germinoma. *J Neuroophthalmol* 2015;35(2):171–4.

The authors report a 25-year-old woman referred for progressive visual loss over 1 year associated with diabetes insipidus. MRI showed a 1.8×1.2 cm enhancing mass of the optic chiasm and pathology of the partial resection revealed a germinoma which are usually located in the suprasellar or pineal location. During embryogenesis germ cells migrate aberrantly into the head or are misplaced within the mesoderm in the region of the optic nerves. The patient was treated with radiation and 1-year follow-up had no detectable tumour on MRI.

Michael Vaphiades

CONVERSION RATE OF OCULAR MYASTHENIA GRAVIS

Nagia L, Lemos J, Abusamra K, Cornblath WT, Eggenberger ER. Prognosis of ocular myasthenia gravis: Retrospective multicenter analysis. *Ophthalmology* 2015 Jul;122(7):1517–21.

The authors conducted a retrospective multicenter study in 158 patients with ocular myasthenia gravis (OMG) to calculate the rate and timing of conversion from OMG to generalized myasthenia gravis. They found that the overall conversion rate to generalized disease was 20.9%. Median time for conversion was 20 months in patients without immunosuppressive treatment, and was 24 months in those who received immunosuppressant.

An-Guor Wang

OPTIC NERVE HYPOPLASIA

Pilat A, Sibley D, McLean RJ, Proudlock FA, Gottlob I. High-resolution imaging of the optic nerve and retina in optic nerve hypoplasia. *Ophthalmology* 2015 Jul;122(7):1330–9.

An observational study was performed to investigate the optic nerve and macular morphology in 16 patients with optic nerve hypoplasia using optical coherence tomography. In addition to the small disc size, they found significantly thinner retinal nerve fiber layer (RNFL), ganglion cell layer (GCL), and inner plexiform layer (IPL) over the nasal macula. They also found signs reminiscent of foveal hypoplasia including continuation of

significantly thicker GCL, IPL, and outer plexiform layer in the central retinal area, with an obvious central doming of the outer nuclear layer.

An-Guor Wang

THE VALIDITY OF ASSESSING PAPILLEDEMA BY OCT

Chang YC, Alperin N, Bagci AM, Lee SH, Rosa PR, Giovanni G, Lam BL. Relationship between optic nerve protrusion measured by OCT and MRI and papilledema severity. *Invest Ophthalmol Vis Sci* 2015 Apr 1;56(4):2297–302.

Since the visual impairment in papilledema is correlated with its severity, it is important to develop an accessible measure of optic nerve protrusion length (NPL) in clinical practice. Optical coherence tomography (OCT) has been used in our daily practice, and its validity of assessing NPL has been examined in this study. The authors found that OCT and MRI measurements of NPL were significantly linearly correlated, supporting the reliability of the OCT-based measurements of NPL in papilledema. In addition, measurements of NPL from OCT and MRI were significantly associated with Frisen papilledema grade. Thus, OCT-based measurement of NPL is a not only sensitive, but also reliable objective measure of papilledema severity.

An-Guor Wang

MITOCHONDRIAL CHANGE IN OPA1 MUTANT

Kao SH, Yen MY, Wang AG, Yeh YL, Lin AL. Changes in mitochondrial morphology and bioenergetics in human lymphoblastoid cells with four novel OPA1 mutations. *Invest Ophthalmol Vis Sci* 2015 Apr 1;56(4):2269–78.

The authors established lymphoblastoid cell lines from four autosomal dominant optic atrophy (ADOA) families harboring different OPA1 mutations. They found increased mitochondrial fragmentation and decreased mitochondrial membrane potential and ATP synthesis in OPA1 mutant cell lines. In addition, there is uncoupled mitochondrial respiration and accumulated oxidative damage. This in-vitro model reveals the OPA1 mutation-induced changes in mitochondrial morphology and bioenergetics.

An-Guor Wang

IS ANTI-VEGF EFFECTIVE FOR AION?

Huang TL, Chang CH, Chang CW, Lin KH, Tsai RK. Efficacy of intravitreal injections of anti-vascular endothelial growth factor agents in a rat model of anterior ischemic optic neuropathy. *Invest Ophthalmol Vis Sci* 2015 Apr 1;56(4):2290–6.

Using a rat model of anterior ischemic optic neuropathy, the authors investigated the efficacy of intravitreal injections of anti-VEGF agents by assessing retinal ganglion cell (RGC) survival and visual-evoked potentials (VEP). They found there is no significant difference in RGC density, apoptosis, and VEP between the anti-VEGF group and control group. The authors conclude that intravitreal injection of anti-VEGF does not have a protective effect in the rat model of anterior ischemic optic neuropathy.

An-Guor Wang

SLEEP APNEA INFLUENCES SECOND EYE INVOLVEMENT OF NAION

Aptel F, Khayi H, Pépin JL, Tamisier R, Levy P, Romanet JP, Chiquet C. Association of nonarteritic ischemic optic neuropathy with obstructive sleep apnea syndrome: Consequences for obstructive sleep apnea screening and treatment. *JAMA Ophthalmol* 2015 Jul 1;133(7):797–804.

The authors examined 118 patients with nonarteritic anterior ischemic optic neuropathy (NAION) for the prevalence of obstructive sleep apnea syndrome (OSAS) and its influence on second eye involvement. 75% (67/89) of NAION patients who underwent polysomnography were found to have OSAS. Increased rate of the second eye involvement at 3 years (15.4% vs. 9.5%) was noted in patients with OSAS, as compared with patients without OSAS.

An-Guor Wang