ABSTRACTS

Neuro-Ophthalmic Literature Review

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OPTIC NERVE SHEATH DIAMETER MEASURED BY ULTRASOUND AT 3MM BEHIND THE OPTIC NERVE HEAD IS REPRODUCIBLE AND ACCURATE

Bäuerle J, Schuchardt F, Schroeder L, Egger K, Weigel M, Harloff A. Reproducibility and accuracy of optic nerve sheath diameter assessment using ultrasound compared to magnetic resonance imaging. *BMC Neurol* 2013;13:187.

The optic nerve sheath diameter (ONSD) is generally increased in patients with increased intracranial pressure and decreased in intracranial hypotension. Therefore, ONSD can be used to estimate intracranial pressure non-invasively. ONSD can be measured by magnetic resonance imaging (MRI) or by ultrasound. MRI provides high spatial resolution, but it is costly and it may not be readily available. On the other hand, orbital ultrasound can be performed with equipment brought to the bedside. How reproducible are ultrasound measurements? In this prospective observation study, the ONSD in right eyes of 15 healthy volunteers was measured by transbulbar ultrasound by two sonographers at two visits 28 ± 11 days apart. These values were compared to ONSD values generated by a 3-Tesla MRI using the coronal images of the HASTE sequence (T2-weighted half Fourier acquisition single-shot turbo spin echo). For each nerve, two measurements were taken: 3 mm and 5 mm behind the optic nerve head. It was found that the ONSD measurements by ultrasound at 3 mm behind the optic nerve head showed good scan-rescan reproducibility and inter- and intra-observer agreement, and were accurate when compared with MRI. However at 5 mm, although the ultrasound measurements were reproducible, they overestimated the ONSD compared to MRI. The authors concluded by recommending sonographic measurements of ONSD at 3 mm behind the papilla as a tool for assessment.

Carmen Chan

CCF IN ASIAN PATIENTS: CLINICAL OUTCOMES

Tan AC, Farooqui S, Li X, Tan YL, Cullen J, Lim W, Leng SL, Looi A, Tow S. Ocular manifestations and the clinical course of carotid cavernous sinus fistulas in Asian patients. *Orbit* 2014;33:45–51.

In this retrospective case series, the authors reviewed the clinical characteristics of 45 patients with carotid cavernous fistula (CCF) confirmed on cerebral angiography who were managed at the Singapore National Eye Centre over a 10-year period. Of the 45 patients, 8 had a direct CCF and 37 had a dural (indirect) CCF; 7/8 of the direct CCFs and 29/37 of the dural CCFs were treated. Of the treated patients, 85% of those with direct CCF and 48% of those with dural CCF had residual symptoms despite treatment. The patients whose dural CCF were treated had a slightly better, but not statistically different, outcome from those who were observed. Poor outcomes were associated with a poor presenting visual acuity ($<6/60$) and use of anticoagulant/antiplatelet agents. This study provides useful prognostic information for those managing Asian patients with CCF.

Carmen Chan

PRIMARY INFLAMMATORY OPTIC NEUROPATHY IN BEHÇET DISEASE

Kidd D. Optic neuropathy in Behçet's syndrome. *J Neurol* 2013;260:7070–7074.

The well-recognised triad of symptoms in Behçet disease consists of recurrent oral aphthosis, genital ulcers, and uveitis. Seventy percent of visual symptoms are related to uveitis, with roughly 30% due to (vaso-occlusive) retinal vasculitis. Only about 10% of patients suffer from neurological complications, such as meningoencephalitis or intracranial venous sinus thrombosis. Afferent visual pathway disruption has been described as a complication of these neurologic sequela, including glaucoma, necrosis of the retina, spread of inflammation backwards from the uveal tract, and intracranial hypertension. In this personal case series and literature review, the author excluded the above complications and presents evidence for the presence of a primary inflammatory optic neuropathy in 20 patients with Behçet disease. In contrast to other forms of inflammatory optic neuropathies, most cases developed subacute, often painless, loss of vision. Recovery of visual function occurred 67% of patients. The only predictor for poor visual recovery was a nadir visual acuity of less than 0.5. Evidence for patient management is class IV (expert opinion only) and consists of prompt administration of corticosteroids and adjunctive immunosuppression in selected cases.

Axel Petzold

TRANSIENT VISUAL SYMPTOMS AS “A FUNNY TURN”

Lavallée PC, Cabrejo L, Labreuche J, Mazighi M, Meseguer E, Guidoux C, Abboud H, Lapergue B, Klein IF, Olivot JM, Sirimarco G, Gonzales-Valcarcel J, Touboul PJ, Amarenco P. Spectrum of transient visual symptoms in a transient ischemic attack cohort. *Stroke* 2013;44:3312–3317.

Funny turn is a term used when a presenting symptom does not point to an obvious diagnosis. This large (2398 patients) French single centre prospective study investigated the spectrum of vision disturbances in patients with transient ischaemic attacks. The prevalence of transient visual symptoms was 34.5%. Patients were divided into six categories: transient monocular blindness (TMB; 36.3%), diplopia (13.4%), homonymous lateral hemianopia (HLH; 12.3%), bilateral positive visual phenomena (phosphenes, photopsias, complex visual hallucinations, palinopsias; 10.8%), lone bilateral blindness (LBB; 4.5%), and unclassifiable (22.6%). The presence of ischaemic lesions was documented by brain imaging (MRI or CT) and complemented by a standardised diagnostic work up. An ischaemic lesion was found in 49 patients (5.9%). These patients were characterised by a longer duration of symptoms (>1 h, mostly HLH) compared with the majority of patients without ischaemic lesions on brain imaging (mostly TMB). Symptomatic stenosis ($\geq 50\%$ or occlusion) was found intracranially in only 2.1% of patients with TMB and extracranially in 12.5% of cases with TMB. An embolic cause was the likely aetiology in 20% of cases (all visual symptoms pooled), most frequently with HLH (atrial fibrillation 23.2%, other source 4.4%) and less so in TMB (atrial fibrillation 5.7%, other source 2.9%). However, it is important to note that “absence of evidence” is not “evidence for absence” regarding embolic aetiology. Overall, the study provides information about transient visual symptoms encountered when evaluating *funny turns*.

Axel Petzold

STENTING TRANSVERSE SINUS STENOSIS IN PSEUDOTUMOR CEREBRI

Radvany MG, Solomon D, Nijjar S, Subramanian PS, Miller NR, Rigamonti D, Blitz A, Gailloud P, Moghekar A. Visual and neurological outcomes following endovascular stenting for pseudotumor cerebri associated with transverse sinus stenosis. *J Neuroophthalmol* 2013;33:117–122.

Transverse sinus stenting for pseudotumor cerebri (PTC) with evidence of venous sinus stenosis is an area of active investigation in the neurosciences. In their report of 12 patients who underwent endovascular stenting, the authors show that the procedure may help with resolution of some but not necessarily all symptoms. Of 88 patients who failed medical therapy, 14 patients with transverse sinus stenosis were identified who met modified Dandy criteria for IIH, had not undergone bariatric or shunt surgery, did not have evidence of another possible cause of IIH, and were not pregnant. Twelve of these patients were found to have a mean pressure gradient of >4 mm Hg, and were subsequently stented. All 12 patients had a significant reduction in the trans-

stenotic pressure gradient. At 6 to 12 weeks after stent placement, visual acuity and visual field (as measured by mean deviation) were stable or improved in 22 of 24 and 19 of 24 eyes, respectively. There was resolution of papilledema in 22 of 24 eyes (grade 0 by Frisén classification). All 11 patients who reported tinnitus had improvement with stenting, including four of seven with bilateral tinnitus who had improvement of symptoms on the contralateral side. Headaches improved or resolved in seven patients, and of the five patients with persistent headaches, all had features consistent with migraine. Four of these five patients responded to migraine medication. Although the study population was relatively small and limited to those who had failed medical therapy, the authors' findings indicate that further investigation is warranted to evaluate the procedure's place in the physician's armamentarium of PTC therapies.

Evan Price

THE 'TIRED EYES' PATIENT MAY HARBOR REAL ABNORMALITIES

Badham SP, Hutchinson CV. Characterising eye movement dysfunction in myalgic encephalomyelitis/chronic fatigue syndrome. *Graefes Arch Clin Exp Ophthalmol* 2013;251:2769–2776.

Chronic fatigue syndrome (CFS) is a prevalent problem causing malaise, generalised pain, and exhaustion. Patients often have depression and complain of cognitive slowing. Visual symptoms are common, but frustratingly amorphous. They include generalised blur, eye pain, and photophobia. This study examines 20 patients with chronic fatigue and compared their eye movements with 20 controls. Saccades, antisaccades, and smooth pursuits were all quantified. The authors determined that while regular saccades did not differ in accuracy between patients and controls, those with CFS had more difficulty with antisaccades. CFS patients also had decreased smooth pursuit gain, and worsened over subsequent trials. This contrasted with controls who performed similarly on all trials. The authors concluded that CFS patients can be tested for objective markers of their visual disturbance, and this may hold the key to understanding this complex syndrome.

John H. Pula

PRIMARY SPONTANEOUS CSF LEAKS AND IDIOPATHIC INTRACRANIAL HYPERTENSION

Pérez MA, Bialer OY, Bruce BB, Newman NJ, Bioussé V. Primary spontaneous cerebrospinal fluid leaks and idiopathic intracranial hypertension. *J Neuroophthalmol* 2013;33:327–334.

The authors report 3 patients and review the literature on idiopathic intracranial hypertension (IIH) and spontaneous cerebrospinal fluid (CSF) leak. These cases and the literature review confirm a definite association between IIH and spontaneous CSF leak based on (1) similar demographics; (2) increased ICP in some patients with spontaneous CSF leak after leak repair; (3) higher rate of leak recurrence in patients with raised ICP; and (4) patients with intracranial hypertension secondary to tumours may develop CSF leak, confirming that raised ICP from other causes than IIH can cause CSF leak. The authors concluded that CSF leak occasionally may keep IIH patients symptom-free; however, classic symptoms and signs of intracranial hypertension may develop after a CSF leak is repaired.

Michael Vaphiades

HOW IMPORTANT IS MRI FOR CHILDREN WITH OPTIC NERVE HYPOPLASIA?

Ramakrishnaiah RH, Shelton JB, Glasier CM, Phillips PH. Reliability of magnetic resonance imaging for the detection of hypopituitarism in children with optic nerve hypoplasia. *Ophthalmology* 2014;121:387–391.

The authors designed a cross-sectional study of 100 children with optic nerve hypoplasia (ONH) who underwent cranial magnetic resonance imaging, physical examination by an endocrinologist, and a standardised panel of

serologic testing in order to identify if they are at risk for developmental delay, seizures, or death. Magnetic resonance imaging disclosed neurohypophyseal abnormalities, including absent pituitary infundibulum, ectopic posterior pituitary bright spot, and absent posterior pituitary bright spot, in 33 children. Magnetic resonance imaging also detected neurohypophyseal abnormalities in 27 of the 28 children with hypopituitarism (sensitivity, 96%). A normal neurohypophysis occurred in 67 of 73 children with normal endocrine function (specificity, 92%). The authors concluded that neurohypophyseal abnormalities on MRI are sensitive and specific indicators of hypopituitarism in children with ONH.

Michael Vaphiades

BLINDNESS FROM GRAVES' CAN REVERSE

Devoto MH, Golnik K, Bernardini FP, Alencar VM. Improvement from no light perception after orbital decompression for Graves' optic neuropathy. *Ophthalmology* 2014;121:431–432.

The authors performed a retrospective review of 3 consecutive patients blind from thyroid-related compressive optic neuropathy treated with transcaruncular medial wall and posterior medial floor orbital decompression. All patients' recovered vision to some degree, some eyes to 20/20. This case series highlights the benefit of this treatment in this patient population with such poor acuity they generally were felt to harbour a very poor visual prognosis.

Michael Vaphiades

CLINICAL FEATURES OF MS ASSOCIATED WITH LEBER HEREDITARY OPTIC NEUROPATHY MTDNA MUTATIONS

Pfeffer G, Burke A, Yu-Wai-Man P, Compston DA, Chinnery PF. Clinical features of MS associated with Leber hereditary optic neuropathy mtDNA mutations. *Neurology* 2013;81:2073–2081.

The authors performed a prospective cohort study and detected 12 cases (with 44 cases identified in the literature) of multiple sclerosis (MS) with Leber hereditary optic neuropathy (LHON). The combined cohort had the following characteristics: multiple episodes of visual loss, predominance in women, and lengthy time interval before the fellow eye is affected, which is very atypical of LHON; conversely, most patients presented without eye pain and had a poor visual prognosis, which is unusual for typical demyelinating optic neuritis. The authors concluded that the co-occurrence of MS and LHON mtDNA mutations is likely to be due to chance, yet the resulting disorder has a distinct phenotype and more aggressive course.

Michael Vaphiades

BEVACIZUMAB IMPROVES VISION IN OPTIC GLIOMAS

Avery RA, Hwang EI, Jakacki RI, Packer RJ. Marked recovery of vision in children with optic pathway gliomas treated with bevacizumab. *JAMA Ophthalmol* 2014;132:111–114.

The authors reported 4 cases of pediatric optic pathway gliomas (OPGs), 2 neurofibromatosis type 1-related and 2 sporadic cases that received treatment with bevacizumab due to progressive visual acuity (VA) or visual field (VF) loss despite prior treatment with chemotherapy or proton-beam radiation. All 4 subjects demonstrated a marked improvement in their VA, VF, or both while receiving bevacizumab-based therapy. Three patients had complete resolution of their VA or VF loss in at least 1 eye (2 of whom had previously received bevacizumab therapy). The authors concluded that bevacizumab should be considered in this patient population in the appropriate clinical context.

Michael Vaphiades

POST-TRAUMATIC AMAUROSIS SECONDARY TO PARAOPHTHALMIC INTERNAL CAROTID ARTERY PSEUDOANEURYSM TREATED WITH PIPELINE EMBOLISATION DEVICE

Kim JD, Barber SM, Diaz OM, Li HK, Jackson RE, Hall D, Lee AG. Post-traumatic amaurosis secondary to paraophthalmic internal carotid artery pseudoaneurysm treated with pipeline embolization device. *J Neuroophthalmol* 2013;33:359–362.

The authors report a 48-year-old woman with acute visual loss in the right eye associated with retrobulbar pain 20 minutes after striking her right temple against a bed post. The right fundus revealed splinter haemorrhages, segmental arteriolar narrowing, optic disc pallor, and macular ischaemia with a prominent cherry red spot in the macula and fluorescein angiography demonstrated delayed choroidal and optic disc perfusion in the right eye. Neuroimaging was consistent with a right ophthalmic artery occlusion from a pseudoaneurysm. A Pipeline embolisation device (PED; Covidien Vascular Therapies, Mansfield, MA) was positioned within the supraclinoid internal carotid artery (ICA) from the level of the anterior choroidal artery extending into the anterior genu of the cavernous ICA. Angiography performed following embolisation demonstrated complete obliteration of the pseudoaneurysm. The patient's vision in the right eye remained light perception 12 months later.

Michael Vaphiades

A “NEW” MACULOPATHY

Abegg M, Dysli M, Wolf S, Kowal J, Dufour P, Zinkernagel M. Microcystic Macular edema: retrograde maculopathy caused by optic neuropathy. *Ophthalmology* 2014;121:142–149.

Wolff B, Basdekidou C, Vasseur V, Mauget-Faysse M, Sahel J, Vignal C. Retinal inner nuclear layer microcystic changes in optic nerve atrophy. *Retina* 2013;33:2133–2138.

After the initial report in 2012 by Gelfand and colleagues describing macular microcystic changes in the inner nuclear layer of patients with multiple sclerosis, similar findings were subsequently reported in other optic neuropathies, notably neuromyelitis optica. The term “macular microcystic edema” (MME), or “oedema” (MMO), was used. The debate on its cause ranges from inflammatory to degenerative. These two new studies add weight to the retrograde trans-synaptic degenerative theory.

Abegg and colleagues performed a retrospective study screening for MMC in 117 consecutive patients (180 eyes) with optic neuropathy of various aetiologies. Causes of optic neuropathy included ischaemia (33%), compression (39%), inflammation (9%), and hereditary (9%). None had multiple sclerosis or neuromyelitis optica, and patients with glaucoma were excluded. The authors noted MME in 16 eyes (8.8%) of 9 patients, a relatively rare finding. MME was associated with younger age and thicker inner nuclear layer. Compared with healthy controls, MME was associated with thinning of the ganglion cell layer and retinal nerve fibre layer.

Wolff and colleagues report a retrospective series of 24 patients (mean age 37 years) with optic atrophy who have MME. Unlike most other reports, this included patients with glaucoma (12 patients), as well as idiopathic optic atrophy, hereditary optic atrophy, multiple sclerosis, trauma, and hydrocephalus. Results also showed an association with retinal nerve fibre layer thinning.

Abegg and colleagues suggest using the term “retrograde maculopathy”—a non-specific effect of optic neuropathy.

Sui Wong

UNDERSTANDING TANZANIAN ENDEMIC OPTIC NEUROPATHY

Kisimbi J, Shalchi Z, Mahroo OA, Mhina C, Sanyiwa AJ, Mabey D, Mohamed M, Plant GT. Macular spectral domain optical coherence tomography findings in Tanzanian endemic optic neuropathy. *Brain* 2013;136:3418–3426.

Approximately 0.3–2.4% of young adults in Dar es Salaam, Tanzania, have bilateral optic neuropathy, also termed Tanzanian endemic optic neuropathy (TEON). The cause of this is not known but thought akin to a form of toxic/nutritional optic neuropathy or Leber's hereditary optic neuropathy, with its characteristic subacute onset,

bilateral, central visual loss. This is the first study to describe the use of optical coherence tomography (OCT) in TEON. The authors report findings in 128 patients over a 2-year period. 250 scans were of sufficient quality for analysis. Severe loss of retinal nerve fibre layer (RNFL) was observed in 185 eyes (74%), at the caeco-central projection ("papillomacular bundle"), with marked sparing of other regions.

Interestingly, microcystic macular changes were observed in 16 (12.5%) patients, bilateral in 9 (7%). These macular changes were associated with more severe retinal nerve fibre layer loss. The authors of this paper also suggest the hypothesis of retrograde trans-synaptic degeneration causing such changes.

Sui Wong