SOCIETY TIDINGS

40th Meeting of the Upper Midwest Neuro-Ophthalmology Group

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The 40th meeting of the Upper Midwest Neuro-Ophthalmology Group (UMNOG) took place on 27 July 2018 in Chicago, Illinois. As usual, it was hosted by Nicholas Volpe, MD and the Department of Ophthalmology of the Northwestern University School of Medicine in its beautiful facilities. John Chen, MD, PhD, Mayo Clinic, Rochester, Minnesota, organized the meeting along with co-chairs Nicholas Volpe, MD, Susan Ksiazek, MD, and treasurer, Peter MacIntosh, MD. Around 60 people were in attendance—ophthalmologists, neurologists, neurosurgeons, and neuroradiologists in academic or community practice, as well as physicians in their residency training and medical students. There were 26 podium presentations that elicited lively audience commentary.

Judy Chen, MD, University of Wisconsin, described a 15-year-old boy who had a diencephalic tumour presumptively diagnosed as a juvenile pilocytic astrocytoma (JPA). But when the tumour grew despite the conventional chemotherapy, a biopsy revealed gangliocytic elements. After partial resection of the mass, the vision declined severely but eventually mildly improved. This case highlighted that JPA can rarely contain gangliocytic features that may alter its behaviour.

Thanujaa Subramanian, MD, University of Wisconsin, presented a case of a patient who rapidly lost vision in both eyes and had some typical clinical features of idiopathic intracranial hypertension and some very atypical features: upgaze deficiency, unilateral lower motor seventh nerve palsy, a high blood leucocyte count, enhancement of the seventh cranial nerve roots, and fresh cerebellar haemorrhages. The debate was whether these atypical features should elicit a non-

idiopathic diagnosis and a more intense evaluation.

John Chen, MD, PhD, the meeting organizer, presented his study of Mayo Clinic data on lumbar puncture opening pressures (OPs) derived from a study of normal aging in 639 subjects residing in Olmsted County, Minnesota, the home of the Mayo Clinic. All lumbar punctures were performed in the lateral decubitus without sedation by Mayo Clinic nurses. None of the patients had papilloedema or any other features to suggest impaired drainage of cerebrospinal fluid. The most important findings were that OPs varied widely between subjects with a 95% reference interval of 82 and 242 mm water. OP was higher than 200 mm H₂O in 15.8% and higher than 250 mm H₂O in 2% of patients. The OP was higher in obese subjects and lower in those aged over 60 years. It was also higher in subjects with obstructive sleep apnea, but not significantly when obesity was factored out except possibly for subjects with OPs greater than 200 mm H₂O. The take-home points were that lumbar puncture OPs, even when performed properly, vary greatly, are higher in obesity and may even exceed 250 mm H₂O in a small fraction of normal subjects.

Tatiana Deveney, MD, University of Michigan, described three patients with leaking arachnoid cysts and pointed out that ophthalmic indicators such as papilloedema and ocular misalignment are important parameters in determining whether surgical drainage of subdural fluid collections or cystoperitoneal shunt is indicated.

Emerson Kendall, DO, Michigan State University, presented a 69-year-old patient with 3–4 transient monocular visual disturbances consisting of seeing bright lights and a dark spot with jagged edges that enlarged. The episodes would

last 15 minutes and were followed by a “dull ache” behind the affected eye. Caregivers naturally attributed this disturbance to migraine. There were no symptoms to suggest giant cell arteritis, and acute phase reactants were not elevated except for a minimally elevated ESR of 53. Temporal artery biopsy showed pathologic evidence of giant cell arteritis! Corticosteroid treatment stopped the episodes and no permanent vision loss occurred. Was this migraine and giant cell arteritis? Was it giant cell arteritis that provoked migraine? Or was it giant cell arteritis with a wildly unusual presentation? Cases like this always arouse extra diligence among caregivers.

Marilyn Kay, MD, University of Wisconsin, presented two patients with paracentral scotomas who had optical coherence tomography (OCT) disclosed abnormalities consistent with acute macular neuroretinopathy. In one patient, the fundus abnormalities were not visible ophthalmoscopically. The cause and treatment of this condition remain unverified. Dr Kay underscored the point that these cases can be mistaken for optic neuropathy and that when there is no afferent pupil defect, OCT becomes critical to diagnosis.

Mangayarkarasi Thandampallayam, MD, Michigan State University, presented a patient with nightblindness (“nyctalopia”) and binocular visual field defects who had undergone multiple bowel resections for Crohn’s disease. Electroretinography (ERG) disclosed marked rod dysfunction. Serum levels of vitamin A were abnormally low, attributable to malabsorption. Vitamin A replacement rapidly reversed the nyctalopia and abnormal ERG. This phenomenon has been reported, but is apparently not widely recognized. Some audience members offered that they had encountered this problem and that it should also be considered in patients with alcoholism and in those who have undergone bariatric surgery.

Robert Tomsak, MD, Wayne State University, presented a series of patients who had clinical elements of “alcoholic” (nutritional) optic neuropathy, but in whom multifocal ERG (mfERG) was also abnormal. Serum folate levels were low. Folate and multivitamin supplements improved vision and convincingly reversed the mfERG in at least one of these patients. Dr Tomsak emphasized that nutritional optic neuropathy should probably be

called as “nutritional optic neuropathy and retinopathy.”

Devon Cohen, MD, Mayo Clinic, reviewed the Mayo Clinic data on 18 patients with high titers of collapsing response mediator protein-5 (CRMP-5) and neuro-ophthalmic abnormalities. The important findings were that 16 of these 18 patients not only had optic neuropathy but also retinopathy, vitritis, or uveitis. In addition, only one patient had enhancement of the optic nerve consistent with optic neuritis. In this cohort, neurologic abnormalities included peripheral neuropathy, ataxia, encephalopathy, chorioathetosis, cognitive impairment, autonomic deficits, myelopathy, and myasthenia gravis. Dr Cohen pointed out that a better term for the condition is “CRMP5-associated vitreoretinopathy and optic neuropathy” as opposed to the current term “CRMP-5 optic neuritis” and that an isolated optic neuropathy would be unusual presentation for CRMP5-associated disease.

Aseem Sharma, MD, a neuroradiologist from Washington University, presented his work on radiologic image processing to improve MRI detection of optic neuritis. His contribution is a post-processing software technique that raises the contrast-to-noise ratio (CNR) and improves conspicuity of subtle imaging abnormalities. Although his technique did not significantly improve detection of contrast enhancement in optic neuritis, it did markedly improve the visibility of the abnormally high T2 signal. Part of the artificial intelligence movement, it will likely become a more prominent component of image interpretation in the future.

David Altshuler, MD, a neurosurgical resident at the University of Michigan, described his study of accuracy in differentiating pituitary adenomas from non-adenomas by MRI interpretation alone. Among two neurosurgeons and two neuroradiologists who viewed scans in a masked fashion, many mistakes were made—equally by both specialists! At least one of those mistakes was in misdiagnosing a sellar aneurysm as a pituitary adenoma, which could have led to disastrous consequences. Dr Altshuler concluded that accuracy could be improved by paying attention to whether the lesion is separable from the pituitary gland (non-adenoma) or inseparable from it (adenoma).

He cautioned that some lesions are so large and destructive that this distinction cannot be made.

Eyal Walter, MD, University of Michigan, reported on 60 patients with skew deviation at his home institution. Skew deviation was nearly always accompanied by other neurologic abnormalities, including nystagmus, ataxia, internuclear ophthalmoplegia, or gaze palsy. The amplitude of the misalignment varied from 1 to 30 prism-diopters (PDs). In patients with two PDs of misalignment, the symptom was blurred vision rather than double vision. The skew deviation resolved in 50% of patients during the study period over an average interval of 8 months. In some others, diplopia could be relieved by prism spectacles or eye muscle surgery.

Aimee Szewka, MD, Rush University, described a patient with several bouts of optic neuropathy in each eye that disclosed areas of papillary elevation on OCT corresponding to new visual field defects. Brain imaging was negative. She asked the audience for thoughts as to the diagnosis. The consensus was that this was recurrent papillitis and that low-dose prednisone and/or azathioprine was a reasonable gesture to try to prevent future episodes.

Amr Sabbagh, a medical student from the University of Michigan, reported a study of 107 inpatients and outpatients with apraclonidine-positive Horner syndrome discovered through a text search of the electronic medical records from that institution between 2001 and 2018. In nearly half of cases, no cause was discovered. Among cases in which a cause was determined, the most common cause was a prior surgical procedure or central venous line placement. Other prominent causes were cervical carotid dissection and cluster headache. Unlike earlier studies, tumour and stroke were minor causes. Comparison of these results to those of earlier studies led to the conclusion that the prevalence of causes of Horner syndrome will vary according to the case accrual method.

Randy Kardon, MD, PhD, University of Iowa, described a new technique called “laser speckle flowgraphy,” which measures the dynamics of choroidal, retinal, and optic nerve blood flow during the cardiac cycle. In arteritic ischaemic optic neuropathy, this technique showed large areas of

reduced choroidal perfusion in the domain of the posterior ciliary arteries, whereas in non-arteritic ischaemic optic neuropathy, it showed decreased perfusion in the border (watershed) zone between the posterior ciliary arteries. Although it may ultimately be clinically useful, it is as yet an expensive research tool with limited availability.

Megan Tuohy, MD, University of Michigan, described two children who began to refuse food in the first year of life and became emaciated. An extensive metabolic evaluation failed to yield an explanation for this life-threatening phenomenon, which was initially attributed to behavioral issues on the part of the children or their parents. Months later, parents noticed ocular oscillations, triggering brain MRI that revealed massive diencephalic tumours, which were pilocytic astrocytomas as part of Russell diencephalic syndrome. Vinca alkaloid-based and platinum-based chemotherapy finally arrested tumour growth and led to the regaining of developmental milestones and disappearance of nystagmus in one child and stabilization of weight loss in the other. Dr Tuohy emphasized the importance of looking for nystagmus, which occurs in most children with this condition. Delayed diagnosis may unfavorably affect outcome, which is often dire.

Chloe Winterbotham, MD, reported patients with non-arteritic ischaemic optic neuropathy in one eye, asking whether it was safe to climb to high altitudes or fly. She summarized the pathophysiology in travel medicine and reviewed a North American Neuro-Ophthalmology Society (NANOS) Net email thread that debated whether mountain climbing or flying was advisable. Alfredo Sadun, MD, PhD had suggested that NAION patients consider flying new, wide-bodied aircrafts with cabin pressure nearer to sea level to reduce hypoxia risk. There was no consensus, but UMNOC audience members generally agreed that there was a rationale for avoiding exposure to low oxygen tension with high altitudes from mountain climbing, but that flying in commercial airlines with pressurized air cabins is not a reported problem.

Anthony Brune, DO, University of Michigan, described three kinds of nystagmus in a patient with multiple sclerosis, abducting nystagmus from

an internuclear ophthalmoplegia, gaze evoked nystagmus, and pendular nystagmus, and pointed out the localization of lesions and the mechanisms believed to generate these patterns.

Kemar Green, DO, Michigan State University, reported a patient with metastatic squamous cell carcinoma who had been treated with pembrolizumab, an immune checkpoint inhibitor whose known adverse side effects include various autoimmune inflammatory conditions. The patient developed diplopia, gait ataxia, ophthalmoplegia, areflexia, and facial diplegia. Ancillary studies confirmed a diagnosis of a Guillain-Barre variant, although ganglioside and standard paraneoplastic antibodies were negative. The neurologic abnormalities rapidly improved after intravenous immunoglobulin treatment. This case calls attention to the need for awareness of the neuro-ophthalmic side-effects of these immunomodulatory agents being increasingly used in the treatment of cancer.

Sangeeta Khanna, MD, St. Louis University, described a patient with diabetic ketoacidosis who underwent rapid correction of blood sugar but developed massive haemorrhagic orbital swelling on both sides, very high intraocular pressures, and profound ischaemia of the extraocular muscles and the eyes. Canthotomy and cantholysis ultimately lowered intraocular pressure, but the patient remained without light perception with mild generalized ophthalmoplegia. MRI showed lack of enhancement of the core of all extraocular muscles. Dr Khanna postulated that the rapid correction of blood glucose played a pathogenetic role but also considered that a catastrophic anti-phospholipid antibody syndrome might have been responsible.

Thomas Mizen, MD, Rush University Medical Center, presented a striking case of

angiostrongyliasis (rat lung worm) that led to a retinal detachment. He showed a remarkable surgical video that revealed the worm under the retina during the retinal detachment repair. It was immobilized with laser and removed from the eye through a vitrectomy port.

Ivy Dreizen, MD, University of Wisconsin, presented two patients without papilloedema but with intermittent severe headaches that improved after lumbar puncture. These cases were discussed by the audience with ultimate determination that one patient likely had IIH while one patient probably did not have IIH.

Peter MacIntosh, MD, University of Illinois at Chicago, described a 19-year-old man with acute, painless, and complete vision loss in the left eye whose vision spontaneously improved to CF in two days. Imaging suggested an idiopathic orbital inflammation or an optic nerve sheath meningioma. There was no response to intravenous corticosteroids and there was a stable appearance on follow-up MRI 2 months later. Should biopsy be pursued given the risk of further vision loss with that procedure? There were arguments on both ways of the debate.

Patricia Davis, MD, Progressive Eye Care, described a 51-year-old woman who underwent endoscopic nasal polypectomy and ethmoidectomy with accidental entry into the right orbit causing enophthalmos and transection of the medial rectus muscle. She used a silicone retina band to reattach the medial rectus muscle through a combined transcaruncular and transconjunctival approach. Postoperatively, the patient had residual enophthalmos and strabismus, but alignment was markedly improved.