Proceedings of the 38th Annual Upper Midwest Neuro-Ophthalmology Group Meeting, July 28, 2017, Chicago, Illinois, USA

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The 38th annual meeting of the Upper Midwest Neuro-Ophthalmology Group (UMNOG) was held on 28th July 2017 at Northwestern University, Chicago, Illinois. There were 53 attendees, the largest number in the history of this meeting. They came from Wisconsin, Minnesota, Missouri, Michigan, Kentucky, Iowa, Illinois, and New Hampshire. Most were neuro-ophthalmologists in academic or community practice, but there were also fellows, residents in ophthalmology or neurology, and medical students. The meeting was organized and chaired by John Chen, MD, PhD, Mayo Clinic, Rochester, Minnesota. The host was Nicholas Volpe, MD, chair of the ophthalmology department at Northwestern University.

The first presentation, delivered by Gregory Kosmorsky, DO, Cleveland Clinic, concerned a patient with unexplained vision loss who had undergone an electroretinogram showing an absent B wave, a clue to dysfunction of bipolar cells. There had been a remote excision of a skin melanoma without known metastases. Dr. Kosmorsky revealed that the patient had antibodies against the TRPM1 receptor, shared by melanoma and bipolar cells. A congenital TRPM1 receptor deficiency occurs in congenital stationary night blindness, another bipolar cell disorder.

Padmaja Sudhakar, MD, University of Kentucky, presented a case of evolving optic neuritis and myelitis with a neuromyelitis optica (NMO) antibody titre that converted from negative to positive. The patient improved dramatically on corticosteroid treatment. The point was to acknowledge that NMO antibody can convert from negative to positive during the course of the illness.

Sahhana Murali, MD, University of Wisconsin, discussed paediatric NMO, pointing out that it is rare in this age group, but that it has more optic neuritis and relapses than the more common adult NMO. Distinction from acute disseminated encephalomyelitis may be difficult.

Talal Derani, MD, University of Michigan, described a case of meningoencephalomyelitis that clinically resembled NMO but had a very high cerebrospinal fluid leukocyte count. NMO antibody was negative. Brain and spinal cord imaging showed features of a venulitis. These imaging features prompted assessment of serum and cerebrospinal fluid antibody to glial fibrillary acidic protein (GFAP), a recently identified IgG antibody found in autoimmune GFAP astrocytopathy. It was positive. Usually associated with cancer, which may be occult, this condition is exquisitely responsive to corticosteroid treatment, as was the case in this patient, in whom no cancer has yet been found.

John Chen, MD, PhD, Mayo Clinic, provided an update on myelin oligodendrocyte glycoprotein (MOG) and aquaporin-4 antibody analysed from stored serum obtained between 1988 and 1991 from 177 patients enrolled in the Optic Neuritis Treatment Trial, a multicentre study conducted across the United States. Only three patients were positive for MOG antibody and none were positive for NMO antibody, indicating that patients in that trial, selected because they had clinically “typical” optic neuritis, generally do not have those antibodies, which are more common in patients with clinically “atypical” optic neuritis. When to order these antibody tests in optic neuritis is still unresolved.

Eric Liao, MD, University of Michigan, described the MRI features of 12 cases of radiation optic neuropathy. He pointed out that the typical imaging feature is a discrete area of enhancement, enlargement, and T2 hyperintensity in the affected prechiasmatic optic nerve. This feature is usually present at the onset of vision loss but may rarely precede vision loss and linger for up to 17 months.

Sophia Chung, MD, St. Louis University, described a 62-year-old woman with acquired comitant esotropia for distance fixation who had no other neurologic manifestations. She pointed out that many terms have been applied to this manifestation, including divergence insufficiency, decompensated esophoria, and, recently, sagging eye syndrome, based on histology showing thinning of the lateral rectus-superior rectus connective tissue bands. She added that acquired comitant esotropia in high myopes may have a different underlying mechanism. In both conditions, base-out or medial rectus recession is usually effective in restoring a wide range of single binocular vision. A neurologic evaluation is not necessary if there are no other neurologic manifestations or a history of cancer.

Jorge Kattah, MD, Illinois Neurological Institute, described a man with a vertebral artery dissection that sequentially impaired perfusion in the anterior and posterior cerebellar arteries, an unusual phenomenon. The issue was whether anticoagulation or antiplatelet aggregation is the proper treatment. He cited literature favouring the latter.

Johanna Beebe, MD, University of Iowa, described a patient with opsoclonus and myoclonus whose spinal fluid was positive for West Nile virus in high titre. Treatment with intravenous corticosteroids exacerbated mania and was replaced with intravenous immunoglobulin. He eventually recovered. Although reported to cause opsoclonus, this association was not known to most members of the audience because it is rare.

Laura Coyne, MD, University of Kentucky, presented a young patient who developed an acute right gaze palsy and internuclear ophthalmoplegia (one-and-a-half syndrome) after bench pressing. Imaging, including digital angiography, disclosed an arteriovenous malformation of the right pontomesencephalic region that had bled. It was treated with stereotactic radiosurgery. Months later, he developed pendular nystagmus together with

synchronous palatal oscillations (oculopalatal tremor). MRI showed the expected hyperintensity of the inferior olivary nucleus, a denervation phenomenon in the Guillain-Mollaret triangle. By eliciting high intrathoracic pressure, bench pressing probably triggers bleeding in intracranial vascular malformations by increasing pressure in draining veins.

Randy Kardon, MD, PhD, University of Iowa, presented the work he had done with Christopher Kirkpatrick, MD, analysing the pupillary responses to topical apraclonidine 0.5% in normal subjects, patients with physiologic anisocoria, and those with Horner syndrome diagnosed by topical cocaine testing, imaging, or other unequivocal clinical findings. They found that reversal of anisocoria is a trustworthy sign of Horner syndrome, but elevation of the upper lid occurs commonly in normal subjects. Dr. Kardon reminded the audience that a positive apraclonidine test results from a lesion anywhere within the sympathetic pathway beginning in the hypothalamus and ending in the third-order neuron emanating from the superior cervical ganglion. He pointed out that other studies have shown that this apraclonidine positivity occurs as early as 48 h after clinical onset of Horner syndrome.

Fernanda Abalem, MD, University of Michigan, presented a patient with subnormal visual acuity in both eyes, bilateral cecentral scotomas, and mildly pale optic discs, who had unequivocally reduced multifocal electroretinogram signals from the region corresponding to the scotomas. The patient had a mitochondrial mutation, proving a diagnosis of Leber Hereditary Optic Neuropathy (LHON). There is only one reported case of ERG evidence of outer retinal dysfunction in LHON, and it is not robust. Dr. Abalem pointed out that mitochondria are present in the outer retina, so this finding is not surprising, although it initially led carers to assign a diagnosis of occult outer retinopathy.

Tatiana Deveney, MD, University of Michigan, reported the clinical outcomes in 17 patients operated for pituitary apoplexy. There were no surgical complications. Recovery from ocular motor cranial nerve palsies and compressive optic neuropathy was excellent and prompt, except in the few patients who arrived with very severe ophthalmoplegia or vision loss.

Susan Benes, MD, formerly from Ohio State University, described the features of patients in whom she conducted neuro-ophthalmic examinations after they had undergone autologous bone marrow-derived stem-cell treatments. She cautioned that the study design was not rigorous and that clinical outcomes were difficult to judge because of the lack of adequate pretreatment documentation and the wide range of pretreatment diagnoses.

Susan Ksiazek, MD, University of Chicago, described a patient who had developed post-operative sequential ischemic optic neuropathy, but who also had intermittent angle closure. She pointed out that ischemic optic neuropathy is, for unknown reasons, not widely documented in angle closure.

Robert Tomsak, MD, Wayne State University, presented a patient who was exposed to high-voltage electricity entering through his forehead. It produced severe binocular vision loss and retinosis. The schisis eventually resolved but severe vision loss persisted. The mechanism was considered to be thermal injury.

Thomas Mizen, MD, Rush Medical College, presented a case of bilateral optic neuropathy caused by infiltration with non-Hodgkin lymphoma. The stirring aspect of this case was that diagnosis was reached by biopsy of the optic nerve head via vitrectomy. (The eye with the biopsied optic nerve was blind.)

Adeela Alizai, MD, South Bend, Indiana, presented a young patient with congenital bicuspid aortic valves and new blurred vision in both eyes weeks after a dental procedure. The patient had been pretreated with antibiotics. Soon he became febrile and weak and began to lose vision. Bedside visual acuity was slightly reduced, but confrontation visual fields and pupils were normal. Ophthalmoscopy disclosed marked optic disc edema with papillary haemorrhages bilaterally. Brain imaging had features consistent with increased intracranial pressure, but lumbar puncture opening pressure was only 24 cm water with normal constituents. Echocardiogram revealed a large vegetation on the aortic valve, as well as an aortic root abscess and severe regurgitation. Emergency aortic valve replacement, abscess debridement, and intravenous antibiotics eventually led to recovery of all systemic and ophthalmic

signs. The optic disc edema was tentatively attributed to increased intracranial pressure not detected on lumbar puncture, perhaps related to congestive heart failure. The case was a reminder that patients may not know that they have bicuspid aortic valves, and that they are at risk for infective endocarditis after even a mild dental procedure.

Michael Lee, MD, University of Minnesota, presented the results of a claims data study, performed in collaboration with the University of Michigan, dealing with the association between patient use of cycline antibiotics and documentation of papilledema or idiopathic intracranial hypertension (IIH). Neither diagnosis was common, and when confounders were excluded, a cause-and-effect association appeared tenuous. Although his team analysed an enormous number of subjects, Dr. Lee acknowledged that such claims data studies have major weaknesses. Even so, he cautioned that clinicians should hesitate before attributing IIH to cycline antibiotic use.

Michelle Abou-Jaoude, MD, University of Kentucky, presented a non-obese woman with features of IIH. She had previously been very overweight and was consuming high doses of vitamin A as part of a weight-loss regimen. Advised to stop taking the vitamins, she was treated with acetazolamide and recovered. Dr. Abou-Jaoude pointed out that IIH can be present in a previously obese patient and that clinicians should be aware that weight-loss regimens may contain vitamin A, which has been implicated in raising intracranial pressure.

Sangeeta Khanna, MD, St. Louis University, presented a patient who met criteria for a diagnosis of IIH, but imaging showed a sagging pons and cerebellar tonsillar descent. She wondered if these signs were secondary to the IIH or represented intracranial hypotension. The patient had not undergone lumbar puncture. The conclusion was that there was inadequate imaging basis for a diagnosis of intracranial hypotension and that there was a caution about performing lumbar puncture, given the slight risk of further tonsillar descent.

Aimee Szewka, MD, Rush University, discussed the causative role of raised intracranial pressure in spontaneous intracranial cerebrospinal leaks. Neuro-ophthalmologists are often consulted to rule out papilledema in such cases. Dr. Szewka

pointed out that even though papilledema is rarely found, these patients may have elevated opening pressures on lumbar puncture. The question is whether patients without papilledema should be placed prophylactically on ICP-lowering medication such as acetazolamide, as is her choice. There was no consensus among the listeners on this point, although this practice seems reasonable when there are repetitive leaks or extensive calvarial remodelling.

These stimulating podium presentations were followed by a fascinating array of 2 min presentations in the final portion of the meeting called “the flea circus.”

Attendees celebrated the largest gathering in the history of this meeting, agreed that they had learned a lot, and resolved to meet again next year—same time, same place.

Anyone interested in attending next year or being included on the UMNOG mailing list should contact Dr. John Chen at chen.john@mayo.edu.

Declaration of interest

The author reports no conflicts of interest. The author alone is responsible for the content and writing of the article.