Proceedings of the Neuro-Ophthalmology Program of the 33rd Asia-Pacific Academy of Ophthalmology Congress, Hong Kong, 8–11 February 2018

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ABSTRACT

The 33rd Asia-Pacific Academy of Ophthalmology (APAO) Congress was held on Feb 8–11, 2018 in Hong Kong. This report summarized the highlights of the neuro-ophthalmology program of the Congress, including the scientific symposia (invited and submitted) and the social activities.

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The 33rd Asia-Pacific Academy of Ophthalmology (APAO) Congress was held on 8–11 February 2018, in conjunction with the 29th Hong Kong (HK) Ophthalmological Symposium at the HK Convention and Exhibition Centre. Over 5200 registered delegates from 63 countries attended the conference.

During this congress, there were six 90-minute invited neuro-ophthalmology symposia organised by the neuro-ophthalmology scientific program committee (Convener: Dr Neil Miller [US]; coordinators: Dr Klara Landau [Switzerland], Dr An-Guor Wang [Chinese Taipei], Dr Yong Zhong [China], and Dr Carmen Chan [HK]; secretary: Dr Andy Cheng [HK]). In addition, there was a paediatric neuro-ophthalmology session organised by the paediatric scientific program committee, a free paper session and 37 posters/e-posters in the neuro-ophthalmology subspecialty.

Invited program

8 February 2018

The neuro-ophthalmology program kicked off with the APAO-ASNOS [Asian Neuro-Ophthalmology Society] Joint Symposium: Challenging Cases in Neuro-Ophthalmology (Afferent), which was chaired

by Drs Anthony Arnold [US], Satoshi Kashii [Japan, President of ASNOS], and Yong Zhong.

Dr Shaoying Tan [China] presented a case of rapidly progressive bilateral sequential optic neuropathy, headache, and weight loss in a diabetic patient. Magnetic resonance imaging (MRI) and computer tomography (CT) showed a heterogeneously enhancing lesion with bone destruction at the orbital apex, which was revealed to be fungal infection with ethmoid sinus mucosal biopsy. The case highlighted the importance of considering fungal disease when visual loss occurs in patients with diabetes mellitus, or who are immunosuppressed by age, disease, or treatment. Dr Clement Tan [Singapore] led the discussion on an unsolved mystery case of bilateral disc swelling, raised intracranial pressure with elevated cerebrospinal fluid protein, peripapillary choroidal neovascularisation/choroiditis, and elevated anti-PR3 antibody. Systemic workup for infection, vasculitis, and malignancy revealed no unifying diagnosis. The patient responded to a combination of oral steroid, acetazolamide, intravenous cyclophosphamide, and intravitreal bevacizumab. Dr François-Xavier Borruat [Switzerland], showed the importance of recognising atypical presentation and causes of toxic optic neuropathy in patients with tuberculosis using two cases. The first case was ethambutol

Figure 1. Photos of some faculty members taken at the end of the Common Neuro-Ophthalmological Scenarios Symposium on 10 February 2018.

Figure 2. Drs Neil Miller and Prem Subramanian received their Achievement Awards from Dr Dennis Lam, scientific program committee chair, at the Gala Dinner on 10 February 2018. Used with permission from the Asia-Pacific Academy of Ophthalmology; photos taken at the 2018 APAO Congress.

toxicity presenting with bitemporal visual field defect. The second patient had bilateral disc swelling which progressed despite stopping ethambutol. Linezolid was identified as the cause. Dr Clare Fraser [Australia] presented a case with a history of a major gastrointestinal surgery suffering from bilateral rapid onset central scotomata with night vision impairment after an episode of severe gastroenteritis. The patient was diagnosed to have retinopathy

secondary to vitamin A deficiency due to his gastrointestinal problems. The role of functional assessment using electrodiagnostic tests was emphasised. Dr Hyosook Ahn [Korea] presented a case of unilateral morning glory disc anomaly. MRI showed thickening of the ipsilateral optic nerve involving the chiasm, which could represent concurrent optic nerve glioma. She was observed, and fortunately the visual functions and MRI findings were

Figure 3. Members of the faculty and friends relaxing at dinner on 9 February 2018.

relatively stable over 10 years of follow-up. Dr Yong Zhong presented a series of cases with visual loss resulting from compressive optic neuropathy due to potentially lethal intracranial lesions masquerading as more benign conditions such as amblyopia, macular disease, and glaucoma.

In the New Concepts in Neuro-Ophthalmology symposium, chaired by Drs Celia Chen [Australia], Shihui Wei [China], and Patrick Yu-Wai-Man [UK], emerging pathophysiological, neuroimaging, and therapeutic concepts in neuro-ophthalmology were reviewed.

Dr Yu-Wai-Man gave us some updates in his symposium “Principles and application of gene therapy and stem cells” focusing on retinal neurodegenerative and mitochondrial diseases. Adeno-associated virus has proven itself to be a very favourable viral vector. Principles of gene therapy for treatment of autosomal dominant and recessive diseases were clearly introduced with therapeutic strategies including correction of gene expression, enhancement of neuronal survival, and allotropic gene expression. As an ideal target organ, several ophthalmological clinical trials are underway as the field of gene therapy is maturing. Dr David Mackey [Australia] showed us a comprehensive approach in the management of hereditary optic nerve disorders. Accurate diagnosis relies on a good family history, exclusion of other organic pathologies, and appropriate DNA testing. Aside from visual rehabilitation, immediate support should include genetic counselling for both the patients and the family. Finally, Dr Mackey focused on recent gene therapy in Leber’s hereditary optic neuropathy which has shown great promise, though its affordability and effectiveness are yet to be determined. In his talk “The role of varicella zoster virus (VZV) in giant cell arteritis (GCA)”, Dr Prem Subramanian [US] presented recent evidence about the discovery of VZV antigens in the temporal arteries of patients with GCA and its relevance in its pathogenesis was critically appraised. With case illustrations, Dr Subramanian highlighted the potential benefit of steroid sparing therapy such as tocilizumab and he emphasised that angiogenesis should not

be inhibited in GCA. Dr Jane Chan [US] reviewed the recent scientific literature supporting glaucoma as a tauopathy and a neurodegenerative disease. These findings will potentially offer future treatment strategies for glaucoma as well as other optic neuropathies. Dr Shihui Wei [China] updated us on the importance in testing myelin oligodendrocyte glycoprotein (MOG) antibody in patients with demyelinating optic neuritis with severe visual loss at onset or relapsing course. MOG-related optic neuritis had distinct clinical features as compared with neuro-myelitis optica-related optic neuritis: MOG-related optic neuropathies tend to have more optic disc oedema and yet a better and faster visual recovery. Dr Anthony Arnold shared his views on the changing paradigms in neuro-ophthalmology imaging. As neuroimaging becomes more accessible, more affordable, less invasive, and more accurate, some of the traditional management logarithms may be changed accordingly. He demonstrated this paradigm shift in the management of Horner’s syndrome, isolated cranial nerve palsy, and transient visual loss. He suggested imaging in all acquired cases of Horner’s syndrome and potentially all isolated third nerve palsy cases. He also emphasised the need for immediate MRI/diffusion-weighted imaging for patients with amaurosis fugax in this new era.

9 February 2018

The Neuro-ophthalmic Emergencies symposium, chaired by Drs Carmen Chan, Neil Miller, and An-Guor Wang, aimed to provide general ophthalmologists with a review of the essential diagnostic points and initial management steps of the life- or vision-threatening neuro-ophthalmic conditions which require prompt treatment.

Dr Karl Golnik [US] started the session with a review of the clinical management of acute Horner syndrome. Although in a recent review of 200 patients with Horner syndrome, about 2/3 were idiopathic and most causes were not urgent; sudden onset Horner’s can be caused by carotid dissection and should be managed emergently. Dr Mahesh Kumar [India]

highlighted the importance of recognising and treating pituitary apoplexy as a medical emergency with three clinical cases. Dr Anthony Arnold explained that idiopathic intracranial hypertension patients could be divided into “not too bad”, “pretty bad”, and “very bad” categories. The “very bad/ fulminant” patient should be managed differently and aggressively, including using lumbar drainage as an emergent treatment. Dr Walter Jay [US] gave us a comprehensive review of the clinical presentation, diagnosis, and treatment of GCA; in particular, the use of a GCA prediction model (available as a smartphone app) and tocilizumab. Dr Dean Cestari [US] highlighted the importance of neuroimaging in most patients with acute onset third nerve palsy. He concluded that CT angiogram was probably better than MR angiogram for detecting cerebral artery aneurysms. Cerebral/catheter angiography carried a low risk of permanent neurological complications and was rarely indicated. Dr Usha Kim [India], an oculoplastic surgeon, concluded the session with a talk on fungal diseases of the orbit, which particularly affect diabetic patients and require a multidisciplinary approach, including prompt surgical debridement, ENT surgeon input, and systemic antifungal treatment.

Paediatric Neuro-Ophthalmology Symposium was one of two scientific sessions during this year's APAO congress with a focus on paediatric neuro-ophthalmic conditions. It was chaired by Drs Jonathan Ho [HK], Jeong-min Hwang [Korea], and Klara Landau [Switzerland, President of the European Neuro-Ophthalmology Society].

Dr Satoshi Kashii gave an overview of neuro-ophthalmologic examination in children. Multiple cases were presented to stress the importance of observation of saccadic and pursuit eye movements, especially in preverbal children. A classification into gaze palsy (divided into supranuclear and nuclear) and strabismus due to peripheral causes was given. The importance of ruling out tumour and trauma in children with apparent “sixth cranial nerve palsy” was highlighted. Dr Patrick Yu-Wai-Man discussed the three common types of congenital optic disc anomalies. Optic nerve hypoplasia (ONH) was one of the three leading

causes of blindness in children. Causes of ONH were inconsistent in the literature, including social deprivation factors, toxin exposure, and maternal type I diabetes mellitus. Disc to macula/disc diameter ratio >4 may be a useful tool to support the diagnosis of ONH. It can be associated with ocular abnormalities including high myopia, nystagmus, and potentially life-threatening condition, including hypothalamic dysfunction and endocrine abnormalities. The other optic disc disorders covered were optic disc coloboma and morning glory anomaly, which can both be associated with choroidal neovascularisation and retinal detachment. Dr Klara Landau gave a talk on swollen optic discs in children. The differentiation of true versus pseudo-disc swelling was prudent and case illustrations highlighted important clinical signs, e.g. tracing of peripapillary vessels and spontaneous venous pulsation. Drusen-like bodies can be present in chronic papilloedema. Investigations might help diagnosis in children including fluorescein angiogram and careful perimetric examination. Dr An-Guor Wang discussed cortical visual loss in children. It encompasses disorders involving malfunction of retrochiasmatic pathways, resulting in visual dysfunction. A review showed 40% of cortical visual impairment (CVI) was due to unknown causes, 23% due to perinatal, and 21% due to genetic or neurodegenerative causes. The latter category was further subdivided anatomically into cortical or deep grey matter lesions, and peripheral or deep white matter lesions. Case examples were presented including hypoxic, traumatic, and mitochondrial causes. History, visual behaviour, ocular, and systemic (particularly skin) features were integral to establish diagnosis of CVI. Dr Jeong-Min Hwang, presented congenital cranial dysinnervation disorders (CCDDs). The talk emphasised the utilisation of high-resolution T2-weighted MRI to detect the presence or absence of third, fourth, and sixth cranial nerves and extraocular muscles. Clinico-radiological studies revealed one-third of congenital third nerve palsy was associated with a hypoplastic or aplastic nerve on MRI. Regarding congenital fourth nerve palsy, absence of nerve on MRI

was seen in 73% and it was associated with hypoplastic superior oblique muscle and early onset head tilt. For Duane syndrome, absence of sixth cranial nerve was seen in most type I and in some type III cases. Absence of both sixth and seventh cranial nerves was seen in Mobius syndrome. Lastly, MRI of the cranial nerves can help to establish the diagnosis of CCDD in atypical presentations. Dr Neil Miller gave the last talk of the session on nystagmus in children. A comprehensive classification including congenital/infantile, acquired causes, and saccadic intrusions was illustrated with salient clinical features highlighted. The differentiation between gaze-evoked versus physiological endpoint nystagmus was discussed. Next, Dr Miller emphasised the importance to rule out sinister causes, such as retinal and optic nerve diseases, which made up almost 1/3 of paediatric nystagmus patients in a review of 71 patients. Also, it is essential to perform a systemic search of neuroblastoma in children presented with ocular flutter and opsoclonus. Treatment options of congenital motor nystagmus including contact lenses, prism, medications, and surgery were discussed.

10 February 2018

The first session of the day was Challenging Cases in Neuro-Ophthalmology (Efferent), chaired by Drs Andy Cheng and Walter Jay.

Dr Hazel Lin, representing Dr Clement Tan [Singapore], discussed a case of a middle-aged lady with multiple vascular risk factors presenting with progressive painful third nerve palsy. The patient was later noted to have aberrant regeneration with lid-gaze synkinesis during recovery. Repeated MRI showed enhancing lesion over the cavernous sinus and with ipsilateral lacrimal gland enlargement. Dr Dean Cestari presented a young gentleman with binocular horizontal diplopia and facial weakness. He had history of medulloblastoma with previous radiotherapy and chemotherapy. Examination of the patient showed right gaze

palsy, right internuclear ophthalmoplegia (one and a half syndrome) with ipsilateral facial paresis. MRI showed an infarct over the facial colliculus. Dr Prem Subramanian discussed a case with systemic dysautonomia of unknown aetiology. The patient presented with Horner's syndrome-like pupil abnormality with dilatation lag and positive response to apraclonidine test in one eye, and Adie's pupil-like abnormality with light near dissociation and positive response to low-dose pilocarpine test in the other eye due to mixed sympathetic and parasympathetic dysfunction. The patient was lost to follow-up so the exact aetiology of the systemic dysautonomia was not elucidated. Dr Sharon Tow [Singapore] shared a patient with ptosis without diplopia or visual complaints. She was also noted to have ipsilateral elevation deficit, prominent superior sulcus, and endophthalmos. CT orbit showed features compatible with silent sinus syndrome or imploding antrum. Dr Andy Cheng shared a series of patients presenting with episodic diplopia. These patients noticed paroxysmal diplopia after brief sustained eccentric gaze. They were finally diagnosed to have ocular neuromyotonia of abducens and oculomotor patterns. Most of these patients received previous radiation therapy for head and neck tumours and many of them showed good response to low-dose carbamazepine. Dr Carmen Chan presented an 11-year-old girl with nystagmus since 6 months of age, with progressive deterioration of vision. The eye movements had features consistent with congenital nystagmus, but she was also noted to have bitemporal hemianopia. MRI and biopsy subsequent reviewed a large inoperable sellar glioma causing compressive optic neuropathy. The importance of further investigations was emphasised when a patient develops alarming symptoms/signs inconsistent with his/her pre-existing clinical diagnosis.

The Common Neuro-Ophthalmological Scenarios Symposium (Figure 1) was chaired by Drs Noel Chan [HK], John Crompton [Australia], and Mahesh Kumar. Dr Crompton shared his experience with encountering

functional visual loss in his paediatric eye practice and emphasized the importance of history taking in the diagnosis to prevent unnecessary investigations and treatment. With his six cases, he also pinpointed the importance of watching out for underlying pathologies such as Leber's hereditary optic neuropathy or sellar masses. Dr François-Xavier Borruat presented a case of bilateral painless visual field constriction with preserved visual acuities and colour vision. Neuroimaging eventually showed bilateral enhancing optic nerves involving the chiasm and the inflammatory optic neuropathy responded to steroid treatment after ruling out infection. Apart from diffused retinopathy, bilateral anterior optic neuropathies, chronic papilloedema, and bilateral occipital lesion, he concluded that differential diagnosis of bilateral concentric visual field loss should also include bilateral posterior optic neuropathies of neoplastic/inflammatory/ infective causes. Dr Walter Jay highlighted some of the recent literature and surveys regarding perioperative visual loss following non-ocular surgery such as cardiac or spine surgery. A recent survey has revealed that perioperative ischemic optic neuropathy in spinal fusion significantly decreased from 1998 to 2012 by about 2.7-fold while the incidence in cardiac surgery did not change. Dr Celia Chen highlighted common differential diagnosis of pseudo-papilloedema and reminded the audience to perform a detailed examination of the disc to look for vascular and mechanical features of true disc swelling. Correlation with optic nerve function and attention to changes in disc appearance over time are crucial as pseudo-oedema and true oedema can coexist in real-life practice. In his talk "Mimickers of retrobulbar optic neuritis", Mahesh Kumar presented five patients who were initially diagnosed as retrobulbar optic neuritis but suffered from intracranial/ sinonasal tumours, tuberculosis arachnoiditis, and Leber's hereditary optic neuropathy. Possible mechanisms of sudden visual loss caused by intracranial mass lesion include ischaemia due to vascular compression or demyelination secondary to compression. Early neuroimaging is important in patients with the

preliminary diagnosis of retrobulbar optic neuritis. Dr Sharon Tow summarised recent literature regarding neuro-myelitis optica-related optic neuritis. She proposed the following criteria for aquaporin-4 antibodies testing in optic neuritis patients: Asian ethnicity, poor presenting visual acuity, altitudinal field defect, no improvement after 3 days of intravenous methylprednisolone, optic nerve enhancement of $\geq 50\%$ length or involving optic chiasm, bilateral involvement, relapsing course, presence of other autoimmune condition, or past history of transverse myelitis.

APSPS [Asia-Pacific Strabismus and Paediatric Ophthalmology Society] Symposium: Paediatric Neuro-Ophthalmology Conditions You Should Not Miss, chaired by Drs Hyosook Ahn, An-Guor Wang, and Wilson Yip [HK], was organised by the paediatric scientific program committee. Dr Craig Donaldson [Australia] started off with the introduction of some of the dangerous strabismus in children. Dr Celia Chen systematically guided the audience through different cases and scenarios in the evaluation of optic disc swelling in her talk "Are the optic nerves really swollen?". Dr Walter Jay summarised recent literature concerning diagnosis and management in traumatic optic neuropathy. Dr Neil Miller gave us a comprehensive review of various optic nerve tumours in children. Dr Ahn presented an interesting paediatric case with iris heterochromia. Dr An-Guor Wang concluded the session sharing a case of a girl with blurred right field.

Submitted program

The Rapid-Fire Neuro-Ophthalmology Update Session was held on 8 February and was chaired by Drs François-Xavier Borruat, Dean Cestari, and Clare Fraser. There were 11 oral presentations, most of them were on the clinical features and treatment of various types of optic neuritis (particularly those related to MOG antibody and neuromyelitis optica); including presentations by Drs Shaoying Tan, Shuo Zhao [China], Huanfen Zhou [China], and Selvakumar Amibika [India]. In addition, there were presentations on laboratory research topics by Drs Bo Young Chun [Korea] and

Hao Kang [China]; non-arteritic ischaemic optic neuropathy by Dr Shan Cao [China]; yield of various diagnostic tests for ocular myasthenia by Dr Akshay Nair [India]; a new visual field screening software by Patcharaporn Jaru-Ampornpan [Thailand]; and a paper by Dr Chaithra Aroor [India] which presented the novel concept of higher ocular aberrations and dysfunctional tear film acting as potential triggers for migraine.

The Best Scientific Paper Award in the neuro-ophthalmology subspecialty, based on submitted abstracts, was awarded for the presentation on “Clinical characteristics and prognosis of paediatric optic neuritis with positive MOG antibodies in China” by Drs Shaoying Tan, Honglu Song, and Shihui Wei [China].

Social program

At the Gala Dinner on 10 February, Drs Neil Miller and Prem Subramanian received Achievement Awards as recognition for their time and contribution to the scientific programs of the APAO Annual Congresses (Figure 2). In addition to the official APAO social program, a dinner was held by the HK hosts for the invited neuro-ophthalmology faculty, on 9 February 2018 at the Golden Valley Restaurant in Happy Valley, HK. About 40 guests attended the dinner (Figure 3). Hopefully all enjoyed the food, wine and company!

The complete program of the APAO 2018 congress and official photos are available from the APAO 2018 website (<http://2018.apaophth.org/>).