CANADIAN NEURO-OPHTHALMOLOGY SOCIETY

THURSDAY 21 JUNE

Paper #0038
Here we go again. A case of recurrent ophthalmoplegia

Mark Gans

Abstract:

A 19 year old male presented with a history of recent onset diplopia. His history was significant for a previous episode of a similar problem some years ago, which spontaneously resolved. There was no other history of note.
A neuro-ophthalmological exam demonstrated a profound bilateral ophthalmoplegia with an otherwise normal eye exam.
An MRI and lumbar puncture were performed leading to a presumptive diagnosis.
The diagnosis in this case and the diffential diagnosis of bilateral ophthalmoplegia will be discussed.

Paper #0048
Tolosa-Hunt Syndrome: a case report and literature review

Adel Al-Buloushi, Abdulmohsen Hussain, Abdulmohsin Jafar

Abstract:

Purpose: Tolosa-Hunt syndrome (THS) has been described as a painful, unilateral, ophthalmoplegia and headache caused by a nonspecific granulomatous process in the cavernous sinus, the superior orbital fissure, or the orbital apex. It is a diagnosis by exclusion.

Methods: A 39 year old healthy Caucasian male presented three week history of a progressive pain of the left eye, headache mainly in the vertex region and cranial nerve palsy of III,IV and VI. Proptosis of 1 mm and ptosis of the left eye were noticed and the rest of the exam was normal. Computed tomography (CT) and Magnetic resonance imaging (MRI) showed opacification of the sphenoid sinus, a lesion in the region of the left cavernous sinus, crowding at the level of the superior orbital fissure and narrowing of the left internal carotid artery in the cavernous sinus segment.

Results a sphenoidotomy via an endoscopic approach was done and a biobsy submitted to pathology which showed chronic inflammation with no evidence of malignancy. Patient was treated with oral prednisolone only. He showed dramatic improvement on follow up visits. Three months later there was a complete recovery of the extraocular movements and diplopia but persistence of some residual ptosis.

Conclusions: Tolosa-Hunt syndrome is a rare disease that should be differentiated from other more frequent causes of headache and ophthalmoplegia, such as sphenoiditis, parasellar tumors, lymphomas, meningiomas, vascular malformations of the posterior communicating artery or intracavernous carotid artery, giant cell temporal arteritis ophthalmoplegic migraine.

Paper #0049
Sphenoid Sinus Mucocele presented as headach and cranial nerve palsy: A case report and literature review

Adel Al-Buloushi, Abdulmohsen Hussain, Abdulmohsin Jafar

Abstract:

A 70 years old male referred from the ENT department complaining of severe headache and diplopia getting worst the past 3 to 4 months .His past medical history includes hypertension. On examination his visual acuity was OD 20/60, OS 20/70 uncorrected. Dilated funduscopy was normal. But there was a restriction of the left eye movement on the lateral gaze

He was diagnosed with left CN VI palsy and a CT scan of the sinus was ordered and a Hess chart done pre-op.

Patient underwent FESS surgery after the CT scan showed a large Mucocele in the sphenoid sinus, where surgery gave him an immediate relief of his headache. Follow up was done and his unilateral CN IV palsy took few weeks to resolve. Patient had follow up for one year post surgery, where he came back complaining of decreased visual acuity OS> OD but no headache, and was found to be only due to cataract only, and CT scan sinus was done with no positive finding. Plus he had a full range of eye movement with a normal Hess chart.

Sphenoid sinus Mucocele in the literature will be reviewed and the role of ophthalmology will be discussed.

Paper #0052
Head Position-Dependent Changes in Ocular Torsion in Skew Deviation

Raymond Buncic, Shuan Dai, Mano Parulekar, Agnes Wong

Abstract:

Purpose: Skew deviation is a vertical strabismus that is often associated with abnormal ocular torsion and head tilt. It has been attributed to imbalance of vestibulo-ocular reflex (VOR) projections from the utricles in the inner ears to ocular motoneurons, but direct evidence is lacking. The utricles lie roughly in the horizontal plane, and they normally detect static positions (tilts) of the head. We postulate that if skew deviation is caused by imbalance of the utriculo-ocular reflex, the abnormal torsion might be head position-dependent. The purpose of this study is to investigate whether ocular torsion differs in upright vs supine position in skew deviation, and to compare the findings in trochlear nerve palsy.

Methods: Patients with skew deviation or trochlear nerve palsy were recruited (n=10 in each group). While sitting upright, double Maddox rod lenses were placed before each eye while the patient fixated on a light. They were instructed to rotate the lenses until the lines became horizontal and parallel. The experiments were repeated in supine position. Changes in torsion (between upright and supine position) were compared between skew deviation and trochlear nerve palsy using ANOVAs.

Results: Skew patients exhibited excyclotorsion in the hypotropic eye in the upright position, and the torsion decreased substantially or disappeared completely in supine position. In contrast, in trochlear nerve palsy, the ocular torsion did not change between upright or supine positions.

Conclusion: The head position-dependent change in ocular torsion provides evidence that skew deviation is caused by imbalance of the utriculo-ocular reflex. The finding that trochlear nerve palsy does not exhibit such change provides a “fourth step” which could be used clinically, in addition to the three-step test: Ocular torsion that differs in upright vs supine position indicates skew deviation, whereas torsion that does not change between upright and supine indicates trochlear nerve palsy.

Paper #0062

Early vs Delayed Surgery for Infantile Esotropia in Human Infants: Effects on Cortical Visual Motion Processing

Christina Gerth, Linda Peckford, Agnes M. Wong, Tom Wright

Abstract:

Purpose: Infantile esotropia is associated with maldevelopment of cortical visual motion processing, as manifested by nasotemporal asymmetry of motion visual evoked potential (motion VEP) responses. The purpose of this study was to determine how early versus delayed repair of infantile esotropia influences the development of motion VEP in human infants.

Methods: Children with infantile esotropia ≥ 40 prism diopter and onset before age 6 months were recruited prospectively. They underwent surgical correction between 4 to 18 months of age. At age 2 years, motion VEPs were elicited during monocular viewing using two oscillating sine wave gratings separated by 90° at 11 Hz. Asymmetric (F1) and symmetric (F2) frequency components were extracted from the motion VEP responses. Two methods were used to determine the presence of a nasotemporal asymmetry: the magnitude of the Asymmetry Index, which was defined as the ratio F1/(F1 + F2) and the phase difference between F1 and F2.

Results: Preliminary data were collected from 10 children who underwent surgery between 6 and 18 months of age. Mean Asymmetry Index was 0.44±0.27 in infants who were surgically aligned by 12 months of age (n=5 and it was 0.74±0.08 in those who were aligned at 12 to 18 months of age (n=5) (t-test, p=0.04). The difference in phase between F1 and F2 was significant different between the two patient groups (t-test, p=0.01).

Conclusions: Early surgery for infantile esotropia may prevent maldevelopment of cortical visual motion processing. More patients are being recruited for this ongoing prospective non-randomized trial.

Paper #0092

Optic Pathway Gliomas and Neurofibromatosis Type 1: Natural History of 39 Patients

Marie-Emmanuelle Dilenge, June Ortenberg, Robert Polomeno, Laura Segal

Abstract:

Background: Children born with Neurofibromatosis type 1 (NF1) have an increased risk of developing Optic Pathway Gliomas (OPGs) during their childhood; this can lead to devastating visual outcomes if not identified and treated in a timely fashion.

Purpose: To describe the natural history of OPGs in patients with NF1.

Methods: Retrospective case series of patients with OPGs and NF1 seen at the Montreal Children's Hospital, where screening imaging is performed on all NF1 patients. Details on patient demographics, tumor location and natural history of the disease were recorded.

Results: The 327 patient database of NF1 patients was reviewed, revealing 39 patients with confirmed OPG (12%), with an average follow-up of 7 years. 20 patients were female (51%), and mean age of presentation of OPG was 6 years, with a total of 14 patients (36%) presenting past the age of 6 years. 7 patients were symptomatic secondary to the OPG (defined as decreased vision or precocious puberty), with 5 of the 7 patients receiving treatment (chemotherapy). These 7 patients all showed chiasmal and/or retrochiasmal tumor in addition to nerve involvement. Final visual outcome was 20/40 or better in both eyes in 33 patients (85%), and the decreased vision was secondary to the OPG in 4 (10%) of the 6 remaining patients.

Conclusion: As postulated by several other recent studies, OPG's can present and progress beyond the preschool years, and children should be screened accordingly. In addition, location of OPG on MRI can be used as a prognostic indicator, with optic chiasm or retrochiasmal involvement of the OPG pointing towards potentially more aggressive tumors (p=.04).

Paper #0121

Rare case report of cavernous sinus syndrome secondary to neutrophilic chemotactic deficiency.

Thomas Lee, Kelvin Leung, James McCabe

Abstract:

Introduction: Neutrophilic chemotactic deficiency is a condition where the host neutrophils are defective in moving to a site of infection and inflammation. This results in a immunocompromised state and an increase propensity for bacterial infections.

Case: A 48 year old male presents with acute onset proptosis preceded by a one week history of left ear pain and three weeks of malaise and symptoms of upper respiratory tract infection. Past medical history was significant for chronic sinusitis requiring numerous surgical procedures. On physical examination, the patient had 20/200 vision bilaterally, bilateral severe chemosis, and proptosis with greater involvement of the right eye than the left eye. Intraocular pressures were elevated at 26 and 24mm Hg for the right and left eye, respectively. The patient had minimal ophthalmoplegia. Fundoscopy showed engorged veins and no papilloedema. Multiple MRI investigation showed bilateral cavernous sinus thrombosis with involvement of the sigmoid sinus. The patient's status worsened and was admitted into ICU for septic shock. Blood culture was positive for atypical gram positive cocci in chains identified as Streptococcus anginosus group bacteria. The acute proptosis resolved over a period of one month with intravenous metronidazole, vancomycin, and cefotaxime. The patient regained 20/20 vision in both eyes, with a return to normal intraocular pressure. Childhood records along with family communications revealed a rare syndrome of neutrophil chemotactic deficiency.

Discussion: This patient represents a rare case of cavernous sinus syndrome as a result of septic thrombosis secondary to neutrophilic chemotactic deficiency and immunodysfunction.

Paper #0144
Temporal arteritis – a dilemma in clinical and pathological diagnosis

Seymour Brownstein, Patrick Gooi, Nigel Rawlings

Abstract:

Purpose: To describe a case of temporal arteritis in which skip lesions resulted in a delay in diagnosis and propose an approach to help minimize this diagnostic clinical and pathological dilemma.

Methods: A 65-year-old woman with suspected temporal arteritis underwent bilateral temporal artery biopsies, which initially were reported by general pathology as negative. Five months later, our ocular pathology laboratory was requested to review the specimens.

Results: After utilizing multiple step sections to identify focal areas of chronic nongranulomatous inflammation of the vessel wall, subsequent serial sections disclosed small skip lesions exhibiting granulomatous inflammation with multinucleated giant cells in the vessel wall bilaterally. The diagnosis of temporal arteritis was augmented with immunohistochemical studies and special stains. From this case and prior experience with this problem, a technique for temporal artery specimen processing was developed to optimize the detection of skip lesions of the temporal artery.

Conclusion: Patients with temporal arteritis may have their corticosteroid therapy prematurely tapered and discontinued because the temporal artery biopsies initially are reported as negative due to skip lesions, which may lead to bilateral blindness.1 We propose that a pathological diagnosis of temporal arteritis should be very carefully investigated for in all cases where there is clinical suspicion, and especially if there are substantial abnormalities in the laboratory analyses (with elevation of the erythrocyte sedimentation rate and C-reactive protein). We recommend the utilization of our described technique of specimen processing and further increasing the number of step and/or serial sections whenever any notable inflammatory cell infiltrate of the vessel wall is demonstrated on histopathological examination.
Reference:
1. Brownstein S. Nicolle DA. Codere F. Bilateral blindness in temporal arteritis with skip areas. Archives of Ophthalmology. 101(3):388-91, 1983 Mar.

Paper #0153
Progressive bilateral ischemic choroidopathy in antiphospholipid syndrome.

Harmeet S. Gill, Agnes M. Wong

Abstract:

Purpose: To report a unique case of bilateral visual scotomata caused by a choroidal infarction secondary to antiphospholipid syndrome.

Methods: The case notes of the patient were reviewed and a literature search was done using MEDLINE.

Results: A 48-year-old man with a plausible history of toxic exposure in the workplace complained of progressive visual field loss bilaterally. Extensive investigations revealed no toxicity. Because of his atypical visual field defect, he was referred to the Neuro-ophthalmology Clinic for evaluation of possible functional visual field loss. On examination, visual acuity was 20/20 OU. Pupils, color vision, anterior segment, and intraocular pressure were normal. Humphrey perimetry revealed a horse-shoe shaped visual field defect that did not respect the vertical or horizontal meridia in the right eye, and a temporal field defect in the left eye. Fundus examination revealed a normal disc and macula with a few patchy areas of retinal pigment epithelial (RPE) mottling and loss in both eyes. Although a full field electroretinogram (ERG) was normal, multifocal ERG revealed abnormal cone responses in both eyes. While waiting for further investigations, the patient developed a stroke and was found to be positive for lupus-like anticoagulant and antiphospholipid antibodies. A subsequent indocyanine green angiography revealed areas of non-perfusion that corresponded with the visual field defect and the patchy areas of RPE loss. The patient was treated with long-term anticoagulation therapy.

Conclusion: Although vaso-occlusive disease involving both the retinal and choroidal circulation is known to be associated with antiphospholipid syndrome, bilateral choroidal infarction has been rarely reported. This case illustrates that choroidal infarction can present with atypical scotomata. Anticoagulation therapy is indicated to prevent further ocular, neurologic and systemic complications.

Paper #0172
Herpes Zoster Ophthalmicus and Contralateral Sixth Nerve Palsy in a Pediatric Patient

Gabriel Chu, Cindy Hutnik, Walter Liao

Abstract:

Case report: Herpes Zoster Ophthalmicus and Sixth Nerve Palsy in a Pediatric Patient
Involvement of the cranial nerves controlling extraocular movement in herpes zoster ophthalmicus (HZO) is rare. There have been several reports of HZV induced *ipsilateral* cranial palsies, but there has been no reports of contralateral cranial nerve palsies in HZO. We report for the first time, a *contralateral* sixth nerve palsy in a pediatric immunocompetent patient with HZO.