

Optociliary Shunts in Multiple Sclerosis

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Introduction: Optociliary shunt vessels develop as a result of chronic retinal venous obstruction. Optic neuritis has never been reported as a causative influence.

Cases: This case series follows two patients with multiple sclerosis from August 1st, 2019 to April 24th, 2024, who developed optociliary shunt vessels after attacks of optic neuritis. A 43-year-old female presented with left visual loss and bilateral superior optociliary shunt vessels. Perimetry showed bilateral peripheral visual field loss. Optical coherence tomography showed bilateral retinal thinning and ganglion cell complex loss. Optical coherence tomography angiography showed reduced capillary density bilaterally. We investigated her and eventually diagnosed her with multiple sclerosis. The second, 49-year-old female, developed right-sided optociliary shunt vessels after an episode of neuroretinitis. Perimetry revealed bilateral central scotomata; optical coherence tomography showed disc and retinal nerve fiber layer edema, and serous retinal detachment; later, ganglion cell complex loss; and reduced capillary density on optical coherence tomography angiography. Neuroimaging revealed demyelination in both, leading to a diagnosis of multiple sclerosis, and therapy was instituted.

Conclusions: We hypothesize, that demyelinating optic neuritis due to multiple sclerosis causes chronic retinal hypoperfusion, leading to subsequent optociliary shunt development in affected eyes. Our case series reveals that eyes with optic neuritis, both previous episodes and fresh cases, can contribute to sufficient retinal vein hypoperfusion to cause the development of optociliary shunts, which should be reported in the literature.

Neuro-ophthalmology

Strabismus Surgery: To Adjust or Not to Adjust?

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Introduction: Adjustable suture strabismus surgery has been practiced now for many decades. For those surgeons who practice and preach this art, they recommend them for all cooperative and willing patients, and even in children and infants. This provides the surgeon a second chance to fine tune his operative results and achieve superior cosmesis and patient satisfaction.

Purpose: Our objective was to assess the long-term success of adjustable strabismus surgery in terms of postoperative alignment.

Methods: We carried out a prospective study utilizing the fornix approach for adjustable strabismus surgery, in mainly horizontal, but also vertical strabismus in adults and cooperative children, to enhance the postoperative outcomes.

Results: This study recruited 50 adults and children with the mean age being 18.34 ± 9.88 years. The mean preoperative horizontal deviation was 48.76 ± 20.35 prism diopters (PD) and the mean postoperative horizontal deviation was 2.73 ± 3.63 PD. The mean preoperative vertical deviation was 4.8 ± 8.54 PD whereas the mean postoperative vertical deviation was 0.86 ± 1.73 PD. The Wilcoxon Signed Ranks test analyzed the difference between the two which was statistically significant ($p = 0.000$). Surgical success, defined as postoperative horizontal alignment within ≤ 10 PD of orthotropia at the end of one year or more of follow-up after surgery, was achieved in 49 (98%) cases. Out of all patients operated via the adjustable suture technique, 35 (70%) of our patients needed suture adjustment.

Conclusion: Adjustable strabismus surgery has very good long-term outcomes in terms of postoperative alignment.

Neuro-ophthalmology

AntiMOG optic neuritis mimicking ION-NA

P. Garcia Robles

79 myopic male who was initially diagnose with NA - ION after presenting to the ER with a sudden visual loss in his left eye, a subtle optic disc edema with a peripapillary hemorrhage and a diffuse visual field defect more dense inferiously

Neuro-ophthalmology

The Impact of Disease-Modifying Therapy on Retinal Thinning in Relapsing-Remitting Multiple Sclerosis

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Purpose: Retinal thinning is a biomarker for neurodegeneration in multiple sclerosis (MS). This study evaluates the effect of disease-modifying therapy (DMT) on retinal nerve fiber layer (RNFL) and ganglion cell-inner plexiform layer (GIPL) thinning over one year in relapsing-remitting MS (RRMS).

Methods: A single-center, prospective study including 80 eyes from 40 RRMS patients. Participants were divided into a No DMT group (N=18) and a DMT group (N=22) receiving glatiramer acetate, ofatumumab, interferon beta-1b, dimethyl fumarate, interferon beta-1a, fingolimod, or ocrelizumab. Optical coherence tomography (OCT) measured RNFL and GIPL thickness at baseline, six months, and twelve months. Disability progression was assessed using the Expanded Disability Status Scale (EDSS). Statistical analyses included T-tests, ANOVA, Spearman correlation, and regression modeling.

Results: The No DMT group had significantly higher RNFL and GIPL thinning rates (p 0.05). Over 12 months, RNFL thinning was 4.03 μm (No DMT) vs. 0.69 μm (DMT), and GIPL thinning was 3.43 μm (No DMT) vs. 2.25 μm (DMT). Correlation analysis showed RNFL/GIPL thickness was negatively associated with EDSS (p 0.05).

Conclusions: DMT slows retinal thinning in RRMS patients, supporting its neuroprotective effect. RNFL and GIPL thickness correlate with disability progression and serve as biomarkers for MS-related neurodegeneration. OCT may aid in treatment decisions.

Financial Disclosure: No

Neuro-ophthalmology

Alice in Wonderland Syndrome

Doctor Eleftheria Filandrianou

Introduction: Alice in Wonderland syndrome (AWS) is a rare condition that causes temporary episodes of distorted perception and disorientation. They're triggered by a dysfunction of temporoparietal junction within the parietal lobe, and the visual pathway, specifically the occipital lobe. This syndrome can affect multiple senses, including vision, touch, and hearing and primarily affects children and young adults.

Methods:Articles, publications and corresponding literature regarding AWS were studied. Comparisons were made between the incidence among the risk groups, and the different manifestations of the condition depending on the age.

Results:Analyzing the studies concluded that almost 30 percent of the people who experienced AWS had infections. Both head trauma and migraines were tied to 6 percent of AWS episodes. But more than half of AWS cases had no known cause. Some possible risk factors include epilepsy, stroke, cough medicine, use of hallucinogenic drugs, brain tumors and stress. Migraine is considered the leading cause for AWS in adults. Infection, usually caused by Epstein-Barr virus considered the primary cause for AWS in children.

Conclusion: AWS may be underdiagnosed. This is because the episodes, which often last only a few seconds or minutes, may not rise to a level of concern for people experiencing them. The fleeting nature of the episodes can also make it difficult for doctors to study AWS and better understand its effects. It is very critical for an early diagnosis, to take a thorough family and patient history, including habits prior procedures and medication.

Neuro-ophthalmology

In Vivo Confocal Microscopy (IVCM): A Double-Edged Sword in Neurodegenerative Disease (NDD) Diagnostics?

Exploring the Potential and Pitfalls of IVCM in Tracking NDD Progression

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Purpose

NDDs pose a growing healthcare challenge, necessitating diagnostic tools for early detection and disease monitoring. IVCM, widely used to study anterior segment pathologies, has emerged as a tool in analysing NDDs as well. This scoping review explores the diagnostic potential of IVCM in tracking NDD progression while critically assessing the controversies involved with its routine clinical integration.

Methods

A scoping review was conducted following PRISMA-ScR guidelines. Studies examining the correlation between corneal nerve parameters—corneal nerve fiber density (CNFD), branch density (CNBD), and fiber length (CNFL)—and NDD progression were analyzed. A literature search was conducted across PubMed and Scopus databases.

Results

IVCM demonstrated optimistic correlations between corneal nerve degeneration and NDD severity. AI-driven analysis has enhanced diagnostic precision, allowing for automated quantification of nerve alterations. However, significant barriers to routine clinical implementation of IVCM remain, including variability in imaging protocols and uncertainties regarding the use of differences in corneal nerve fibre parameters to distinguish between a diverse set of NDDs.

Conclusions

Alone, IVCM holds promise as a non-invasive, accessible tool for NDD diagnostics, but its widespread clinical adoption is hindered by methodological inconsistencies and unresolved questions regarding its ability to distinguish between NDD pathologies. Future research should focus on standardizing imaging protocols, validating AI-driven quantification methods, and conducting longitudinal studies to establish specific corneal nerve parameters as reliable biomarkers for NDD progression. While IVCM may offer a novel avenue for NDD evaluation, its role as a standalone diagnostic tool remains controversial.

Neuro-ophthalmology

Fulminant Intracranial Hypertension: The New Role of the Ophthalmologist

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Objectives

To review the management of Idiopathic Intracranial Hypertension (IIH) with a risk of progression to a fulminant form.

Material and Methods

Case report.

Results

We present the case of a 31-year-old woman who arrived at our emergency department with a prior diagnosis of IIH from the referring hospital. Upon admission, she met the modified Dandy criteria and had a visual acuity of 20/20 in both eyes (OU). She exhibited severe dyschromatopsia, concentric reduction in 30-2 computerized perimetry, and retinal ganglion cell (RGC) atrophy in both eyes, along with bilateral grade IV-V Frisen papilledema. The patient underwent maximum medical treatment, lumbar punctures, and a ventriculoperitoneal shunt (VPS), yet she progressed to bilateral and permanent vision loss within three weeks.

Conclusions

Certain risk factors in IIH may predispose patients to fulminant progression. Identifying these factors at diagnosis is a criterion for intensifying medical and surgical treatment. Among the surgical options, optic nerve sheath fenestration is considered the most evidence-based technique currently available, although access remains limited. Fulminant forms of IIH account for no more than 3% of cases, and no unified criteria exist for defining them. However, there is consensus in classifying them as cases with severe and rapid visual loss (within four weeks from symptom onset) that progresses over days. Given the devastating consequences in terms of visual function, we emphasize the importance of staging and personalized multidisciplinary management of IIH, as well as the early referral of high-risk patients to a tertiary care center.

Neuro-ophthalmology

THRIVE Phase 3 Study in Active Thyroid Eye Disease: Efficacy and Safety at 15 Weeks of Veligrotug (VRDN-001), a Full Antagonist Humanized Monoclonal Antibody to IGF-1R

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Introduction:

Efficacy and safety data for veligrotug vs placebo at 15 weeks (primary timepoint) were evaluated from an ongoing phase 3 RCT in patients with active TED (THRIVE, NCT05176639).

Methods:

Adults with moderate-to-severe active TED (onset ≤ 15 months, proptosis ≥ 3 mm, and clinical activity score [CAS] ≥ 3) were randomized to 5 IV infusions Q3W of 10 mg/kg veligrotug or placebo. Efficacy outcomes and treatment-emergent AEs (TEAEs) were assessed through 15 weeks.

Results:

113 patients received veligrotug (n=75) or placebo (n=38). Baseline values for veligrotug vs placebo were balanced: mean proptosis, 23.2 mm in each group; CAS, 4.5 vs 4.8; diplopia, 67% vs 68%. 15-week results for veligrotug vs placebo were as follows: proptosis responder rate (PRR; ≥ 2 -mm reduction vs baseline by Hertel exophthalmometry), 70% vs 5% (p0.0001), mean reduction of 2.9 mm vs 0.5 mm (p0.0001); PRR by MRI/CT, 69% vs 9% (p0.0001), mean reduction of 2.9 mm vs 0.6 mm (p0.0001); mean CAS reduction of 3.4 vs 1.7 (p0.0001). In patients reporting subjective diplopia on the Gorman subjective diplopia scale at baseline, complete resolution was reported by 54% vs 12% (p0.0001). TEAEs occurred in 88% vs 63%, most being mild; 4 patients (veligrotug) had serious TEAEs (all unrelated to treatment). Hearing impairment TEAEs occurred in 16% vs 11%.

Conclusions:

Topline results from the THRIVE phase 3 RCT show 5 IV infusions of 10 mg/kg veligrotug were well tolerated and led to significant and clinically meaningful improvements in TED signs/symptoms at 15 weeks. Additional follow-up through 52 weeks is ongoing.

Neuro-ophthalmology

THRIVE-2 Phase 3 Study in Chronic Thyroid Eye Disease: Efficacy and Safety at 15 Weeks of Veligrotug (VRDN-001), a Full Antagonist Humanized Monoclonal Antibody to IGF-1R

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Introduction:

Efficacy and safety data for veligrotug vs placebo at 15 weeks (primary timepoint) were evaluated from an ongoing phase 3 RCT in patients with chronic TED (THRIVE-2, NCT06021054).

Methods:

Adults with moderate-to-severe chronic TED (onset 15 months, proptosis ≥ 3 mm, any clinical activity score [CAS]) were randomized to 5 IV infusions Q3W of 10 mg/kg veligrotug or placebo. Efficacy outcomes and treatment-emergent AEs (TEAEs) were assessed through 15 weeks.

Results:

188 patients received veligrotug (n=125) or placebo (n=63). Baseline values for veligrotug vs placebo were balanced: CAS ≥ 3 , 57% vs 52%; diplopia, 52% vs 59%. 15-week outcomes for veligrotug vs placebo were as follows: proptosis responder rate (PRR; ≥ 2 -mm reduction vs baseline by Hertel exophthalmometry), 56% vs 8% (p0.0001), mean reduction of 2.34 mm vs 0.46 mm (p0.0001); PRR by MRI/CT, 48% vs 3% (p0.0001); overall responder rate (PRR and no worsening in CAS), 56% vs 7% (p0.0001). In patients reporting subjective diplopia on the Gorman subjective diplopia scale at baseline, diplopia responder rate was 56% vs 25% (p=0.0006) and complete resolution was reported by 32% vs 14% (p=0.0152). Most TEAEs were mild; most common was muscle spasms (36% vs 6%). Hearing impairment TEAEs occurred in 13% vs 3% and serious TEAEs in 2% vs 3% (1 related to treatment in each group).

Conclusions:

THRIVE-2, assessing veligrotug in chronic TED, is the first RCT to show statistically significant improvement not only in proptosis but also in diplopia, with a generally well-tolerated safety profile. Follow-up through 52 weeks is ongoing.

Neuro-ophthalmology

Improvement in visual acuity after treatment of optic nerve glioma with Selumetinib in neurofibromatosis type 1

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Purpose: To present a case of a 5-year-old girl with a glioma related to neurofibromatosis type 1 and optic pathway involvement, who after failed treatment with chemotherapy, was successfully treated with Selumetinib.

Methods: A single case report.

Results: A 5-year-old girl with a genetic diagnosis of neurofibromatosis type 1 was referred due to findings of bilateral hypothalamic-optochiasmatic glioma on brain MRI. Ophthalmologic evaluation revealed best-corrected visual acuity of 0.5 in the right eye and 0.3 in the left, along with a temporal atrophy and nasal involvement in the nerve fiber layer (RNFL) optical coherence tomography of both eyes. Due to the findings, chemotherapy treatment with Vinblastine was started. However, despite achieving stability of the glioma on MRI, the loss of visual acuity continued to progress until 0.2 in the right eye and 0.1 in the left eye, as well as the involvement of the RNFL and the visual field. Due to worsening vision, it was decided to switch to a targeted therapy with Selumetinib. After 5 months, a 50% decrease in the size of the glioma was detected. Despite not being able to reverse the atrophy of the RNFL, the visual field and acuity showed a great improvement.

Conclusion: Selumetinib is showing good results in the treatment of unresectable gliomas affecting the optic pathway, not only in imaging results, but also in visual outcomes. This can represent a promising approach to be an alternative to standard chemotherapy and even as a first-line therapy.

Neuro-ophthalmology

Unmasking prostate cancer through ophthalmic herpes zoster: a case report

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Purpose

To describe the discovery of advanced prostate cancer in a patient as a result of severe ophthalmic herpes zoster, as well as the follow-up of his ocular involvement.

Methods

A 67-year-old man was admitted with ophthalmic herpes zoster affecting the right hemiface and associated preseptal cellulitis. Initial examination revealed proptosis of the right eye, complete paralysis of the right third and fourth cranial nerve and a non-reactive mydriasis. Visual acuity (VA) was limited to finger counting at one meter.

Over a month, the condition showed a torpid progression despite systemic treatment. Suddenly, a coincidental finding of a pulmonary infiltrate prompted a thoracic CT scan.

Results

CT scan revealed blastic vertebral lesions, suggestive of metastatic involvement, most likely of prostatic origin, which was confirmed by a significantly elevated PSA level (1911) and a prostate biopsy.

Thanks to intravenous antiviral treatment and corticosteroids, skin lesions and cellulitis improved within a few weeks. However, the patient still exhibited paralysis of the third and fourth cranial nerves, corneal hypoesthesia, and non-reactive mydriasis. The only ophthalmic treatment administered was Alphagan 2 times a day.

Three months later, ocular motility was almost recovered, with only minimal residual mydriasis. VA in the right eye improved to 0.5, demonstrating significant overall recovery.

Conclusions

When faced with an aggressive and slow-evolving ophthalmic herpes zoster, we must suspect the presence of an underlying disease causing immunosuppression.

With antiviral and corticosteroid treatment, skin lesions improve within a few weeks, although ophthalmological involvement may persist for several months.

Financial Disclosure: No

Neuro-ophthalmology

Orbital Cellulitis in a Patient with Sinusitis and Previous Cocaine Use: A Case Report

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Purpose:

This case report shows a rare case of orbital cellulitis in a 49-year-old woman with a history of maxillary sinusitis and past cocaine use. The case highlights the diagnostic and therapeutic challenges associated with cocaine-induced bone destruction and its implications in orbital infections.

Methods:

A 49-year-old woman presented with severe ocular pain, nausea, vomiting, and binocular diplopia. Her medical history included maxillary sinusitis treated with ciprofloxacin and amoxicillin, hypertension, anxiety-depression, and cocaine use.

On ophthalmologic examination, she exhibited complete paralysis of extraocular movements (EOMs) in the right eye, mild palpebral edema, and temporal chemosis. Laboratory findings revealed leukocytosis and elevated C-reactive protein (CRP).

A computed tomography (CT) scan demonstrated phlegmonous changes in the right orbit and bone destruction communicating with the nasal cavity, attributed to cocaine use. No evidence of venous thrombosis was observed.

Results:

The patient was diagnosed with right orbital cellulitis and received intravenous ceftolozane/tazobactam, followed by tazobactam and linezolid for six weeks. She showed progressive clinical improvement, with a reduction in orbital inflammation confirmed by radiological follow-up. Due to the favorable outcome, no surgical intervention was required.

Conclusions:

This case underscores the importance of recognizing orbital cellulitis in patients with sinusitis and drug use. Early broad-spectrum antibiotic therapy and close monitoring were key to successful management, preventing complications and avoiding surgery. A multidisciplinary approach is essential for complex orbital infections.

Financial disclosure: None