Aim of Today’s Lecture

1. Focus on N-Oph dz where neuroimaging recently advanced our understanding of underlying pathophysiology, diagnosis and treatment
2. Emphasize, how imaging helps N-Oph and Neuro-Oph helps imaging in differential diagnosis
   - We will discuss:
     - Pseudotumor Cerebri Syndrome (PTCS)
     - Intracranial Hypotension
     - Leptomeningeal disease
     - Optic neuritis (ON)

Definitions

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<th>Risk Factors for the Pseudotumor Cerebri Syndrome</th>
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Other neuroimaging findings in PTCS


Fulminant idiopathic intracranial hypertension

**PTCS associated with giant arachnoid granulation**

**Therapy: Guided by Neuroimaging Findings & Symptoms**

Case of 47 y/o WM

- WITH RECENT RIGHT UPPER ROOT CANAL
- NEXT DAY DEVELOPS SEVERE H/A WHILE DRIVING X 8 HOURS
- H/A IS AT BASE OF SKULL WITH FEELING OF OFF BALANCE WHEN MOVING HEAD SIDE-TO-SIDE; BLURRED VISION with sudden head motion OU
- NO PMH; NO FH OF MIGRAINE; NO TRAUMA

Diagnosis?
DDx of meningeal enhancement
1. Chemical “meningitis” (chemotherapeutics, heavy metals)
2. Granulomatous infiltration (sarcoid, tb)
3. Inflammation (RAs, eosinophilic granuloma)
4. Infection (viral, bacterial, fungal)
5. Neoplastic “meningitis” (carcinomatous, hem)
6. Subarachnoid haemorrhage
7. Trauma, intracranial surgery
8. Venous thrombosis

Neuro-ophthalmologic Complications in the Patient with Cancer
- Tumor cells reach the subarachnoid space either through the blood, by growing along nerve & vascular sheaths, or by migration from a tumor adjacent to CSF (parenchymal, bony lesions in the skull or spine)
- Katz et al. reported not only ONS coverage with tumor cells but neoplastic invasion along the Virchow-Robin spaces (mesenchymal septae) resulting in demyelination, and axonal beading and degradation of the optic nerve itself.

Germinoma
- 80-90% along IIIrd ventricle
  - 5-10% basal ggl
  - Bifocal germinoma: pineal + suprasellar
- DWI restriction: high cellularity
- Intense homogeneous Gad+
  - ± CSF seeding
  - ± brain invasion
• Cardiac transplantation on immunosuppressants
• CN V, VI followed by complete ophthalmplegia;
• LR atrophy

MRI in Optic Neuritis
MS vs. NMO
➢ Abnormal optic nerve enhancement in 94% of affected nerves (Kupersmith et al.)
➢ ON hyperintensity on FLAIR in 82-100 %
➢ Abnl. signal length >17.5 mm and canalicular location a/w poor or slow recovery from ON even if treated with steroids
➢ Simultaneous ON OU in a monocularly symptomatic patient or chiasmal enhancement should warrant careful evaluation for NMO!

Optic tract lesion
First sign of MS

1 year later facial and upper extremity numbness, bilaterally

Demyelinating plaques

40 Y WF pain on EOM x 1 wk, abrupt onset ↓ VA: CF 1’ OS

Enhancement
may persist up to 4 mo
non-specific marker of breakdown of BBB
intraorbital & canalicular segment
Lesion length > 17.5 mm – poorer prog.
MRI & Eye as Predictors of MS

- MRI: best predictor of present and future risk of CDMS not for diagnosis in 5-yr: 16 vs 51%; in 10-yr 22 vs 56% if no vs 1 or > brain lesion(s); in 15-yr 25 vs 72%
- After ON highest risk of developing MS: in 5 yrs;
  If baseline MRI- then ↑risk if preceding viral syn.
- If no eye pain or NLP or severe disc edema or peripapillary hem. or macular exudates then No MS
- Revised McDonald criteria: Dx of MS with MRI can be made at initial presentation

Where OCT & MRI meets: Pathophysiology

- Focal demyelinated plaques: varying levels of inflammation, gliosis, neurodegeneration
- Evidence: permanent disability correlates best with↓CNS neurons & axons not demetylination!
  Infer from OCT: structural info of retina & ON
- Thinning of RNFL & ↓MV found in MS pts, both with & w/o distinct episodes of ON suggesting ongoing neuronal & axonal loss
- OCT: Macular thinning predominant phenotype

Optical Coherence Tomography (OCT)

- Presenting with ON OS 1 year earlier. She had no history of ON OD.
  Fundus: optic disc palsy OD. SD-OCT demonstrates RNFL thinning in both eyes, suggesting prior subclinical optic nerve damage in OS and optic nerve damage in OS secondary to known ONL.

Neuromyelitis optica Devic's disease NMO spectrum disorders

- Severe inflammation & necrosis of ONs & spinal cord (>5 yrs 50% is blind in 1 or both eyes or need walking aid)
- Non-caucasian, mean age: 40 yrs, ♀:♂=9:1
- 80% +Anti-NMO-IgG serum Abs
- A/w other autoimmune dz: celiac, MG & systemic infection: hepatitis, Lyme, syphilis, TB
- Poor visual recovery= bilat. ON (NMO), LETM, intereye RNFL asymm. >15μm (>3 mths of ON)
- On low immunosupp: Ocular toxo, CMV retinitis (important DDx of visual loss in NMO)
- Beta-IFN for MS considered harmful in NMO!
Manifests significant optic disc pallor in OS and marked intereye asymmetry (43 μm) of the mean RNFL thickness, reflecting significant optic nerve damage in OS. His VA was 20/20 OD, and 20/100 OS, with a left RAPD, a dense central scotoma in the left visual field, and absent color vision in OS.

In eyes with ON both TD- & SD-OCT showed thinner RNFL in NMO vs RR MS.

Homonymous Hemimacular Thinning: A Unique Presentation of Optic Tract Injury in Neuromyelitis Optica

Romero, Rebecca S.; Gutierrez, Ismael; Wang, Eugene; Reder, Anthony T.; Bhatti, M. Tariq; Bernard, Jacqueline T.; Javed, Adil

doi: 10.1097/WNO.0b013e3182504688

DDx of Optic Neuritis & abnl. MRI

- Isolated inflammatory: MS, NMO, ADEM, AION, CRION, postvaccination ON, anti-myelin oligodendrocyte glycoprotein (MOG)-associated Diagnosis of exclusion, recurrent events, worsening with steroid withdrawal
- Systemic dz. associated eg. GCA, paraneoplastic, sarcoid, SLE, Wegener granulomatosis
- Infectious: B. h neuroretinitis, CMV, Lyme, mycosis, syphilis, TB, WNV

Case of 38 y/o AA

- Neurosarcoïdosis
- abnl. CXR in >90% of NS
- WM small vessel vasculitis/angiitis
- Coats CNs/fill IAC

- Leptomeningeal dz of the base of the brain
- spreads along the Virchow-Robin spaces to form intraparenchymal masses
Neurosarcoidosis

45 Y/O WOMAN WITH CONFUSION

Solitary or multifocal CNS masses (non-caseating granulomas)

50 Y/O WW

Eye pain worse with movement

Patient presents with severe periorbital or retro-orbital pain of acute onset that is constant in nature. Diplopia due to ophthalmoparesis usually follows the onset of the pain. The symptoms are usually unilateral.

Strep and WNV meningoencephalitis

Relapsing-remitting clinical course

46 Y/O AA

- Acute visual loss OU;
- Few weeks earlier brain cryptococcus infection
Thank you!