Neuro-ophthalmology Webinar
22 October 2022

Responses to questions raised

Misha Pless questions

- What could be a reason for optic neuritis with papilledema in a 11 year old girl (Indian ethnicity), no other abnormalities found in MRI (no MS, NMO), CSF, no antibodies (AQP4 and Anti-MOG), after steroid i.v. vision again to 6/6, then 2 days after stopping steroids, the other eye developed an optic neuritis, again no cause found, after steroids i.v., I saw the girl today and vision recovered again to 6/6. Otherwise healthy girl. Any recommendations for follow-up? Thank you!
- If liver function is altered which immunosuppressants are better in case of NMO and MOG related optic neuritis?
- Up to which level of altered liver function we can continue Azathioprine in NMO and MOG related optic neuritis?

The differential diagnosis includes atypical optic neuritis not only because of the rapid response to steroids but also the presentation of optic neuritis in the fellow eye after completion of the steroid treatment, underscoring the importance of distinguishing demyelinating optic neuritis from atypical optic neuritis. The causes are different. So are the treatments. Autoimmune optic neuritis, including MOG-AD, still belongs in the differential diagnosis since it could be that the patient has not developed the antibodies in measurable amount yet or the incorrect assay was used (ie, ELISA instead of cell-based assays, etc) for detection. However, ADEM would still need to be considered in a child in this age group. These cases often turn out to be MS in the future analysis. TB would also need to be considered in an Indian girl. Also, very important to ask oneself why liver function is a problem instead of hastily considering immunosuppression. Many causes of systemic disease cause liver involvement and optic neuritis.

Andrew Lee questions

- For the first two speakers, what is the chapter/book/article that covers best in your opinion the subject you just presented?
- Great lectures! I have a question for dr. Lee. What would be you approach in a child with ophthalmoplegic migraine? Any medical treatment?

Ophthalmoplegic migraine is now called recurrent painful ophthalmoplegic neuropathy (RPON).

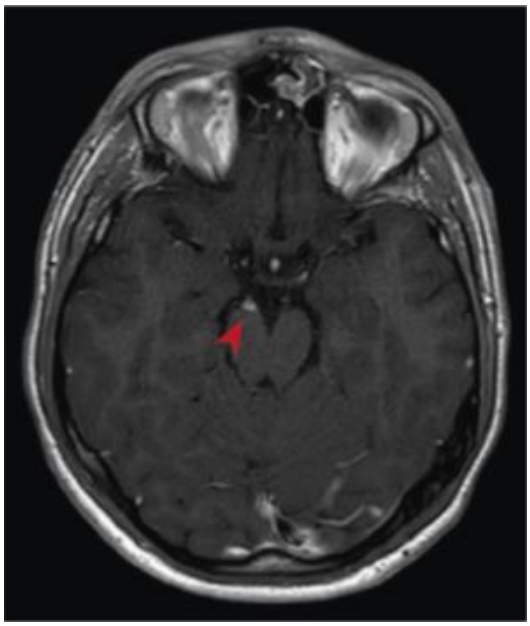
The International Classification of Headache Disorders 3rd edition-beta version (ICHD3β) defined the following diagnostic criteria for RPON:

1) at least two attacks

2) unilateral headache accompanied by ipsilateral paresis of one, two, or three ocular motor nerves
3) the exclusion of orbital, parasellar, or posterior fossa lesions by an appropriate investigation
4) the absence of another diagnosis that could better account for the patient's condition

On the first attack you cannot make the dx/o RPON yet and thus a "full court press" work up should be performed (complete history and exam, MRI head/orbit with gadolinium, LP, indicated labs (usual suspects).

On the second attack, although technically the criteria are fulfilled we should consider repeat imaging but the distinctive sign is enhancement of the nerve in the cisternal segment.



Steroids can be given in the acute phase.

Migraine prophylaxis and treatment could also be considered even though the ICHD moved RPON out of migraine classification.

Jonathon Trobe questions

What might be a safe approach when we see just a no cup disc?
No evaluation needed unless you believe that the optic disc represents ACQUIRED ELEVATION and patient has new visual symptoms and evidence of visual dysfunction or some suggestion of an intracranial process.

How about bilateral nutritional papillary edema?
O*ptic disc swelling is unusual in "nutritional deprivation." It can be seen rarely in Wernicke disease, which you might suspect if the patient is alcoholic or has undergone recent bariatric surgery.*

If there is no family history, which are your Symptoms & signs to suspect LHON? How do you place it in your work up?
Subacute painless vision loss in one eye attributable to optic neuropathy in a young man with peripapillary optic disc nerve fiber layer swelling +/- peripapillary hemorrhage +/- ipsilateral afferent pupil defect and central or cecocentral visual field defect.

What is your opinion in the use of ultrasound to diagnose and follow up papilledema by intracranial hyoertension?
Ultrasound is not useful in this setting. Use visual fields, patient symptoms, and optical coherence tomography.

How do you treat nonarteritic ischemic neuropathy?
There is no effective treatment. Your attention should be directed toward prevention of the same event in the unaffected eye: principally to avoid nocturnal hypotension and to reduce the conventional arteriosclerotic risk factors (diabetes, uncontrolled systemic hypertension, obesity, smoking, dyslipidemia). Overtreated systemic hypertension is a major factor that is not acknowledged by general physicians and internists, who often aim to keep blood pressure low to protect the heart and kidneys. That is a worthwhile aim, but it places the unaffected eye at risk.

What could be a reason for optic neuritis with papilledema in a 11 year old girl (indian ethnicity), no other abnormalities found in MRI (no MS, NMO), CSF, no antibodies (AQP4 and Anti-MOG), after steroid i.v. vision again to 6/6, then 2 days after stopping steroids, the other eye developed an optic neuritis, again no cause found, after steroids i.v., i saw the girl today and vision recovered again to 6/6. Otherwise healthy girl. Any recommendations for follow-up? Thank you!
You are describing recurrent "papillitis" of unknown cause, a condition often encountered by neuro-ophthalmologists. If you have excluded such conditions as sarcoidosis, IgG4 disease, lymphoma, and syphilis, you can probably call this "idiopathic," which is a common occurrence. I often place such patients on low-dose prednisone (5-7.5mg/day) as a preventive gesture, although there is no evidence to support that measure. Be sure you have done enough lab work and imaging to exclude the above-mentioned conditions.

Karl Golnik questions

1. Did someone of the panel notice a raise in GCA during the covid period? can covid infection trigger GCA? I have noticed a raise in bilateral visual loss due to GCA during this period
   I have not noted a rise in GCA patients since the pandemic. If anything, I have seen less.

2. Prof Karl Golnik thank you for your presentation, we had several patients with covid 19 and optic neuritis how do you determined the pathogenesis whether this is because of inflammation or vascular related because some of our patients had hypercoagulable state? thank you.
   I don't think one can determine the pathogenesis for certain but inflammatory optic neuritis typically causes enhancement of the optic nerve on MRI whereas ischemia does not.

3. What line should we follow when asked to examine a patient in ICU?
   I am not sure what this question means - what is a "line" - the protective equipment? I would use whatever is suggested at their institution.

4. Do you have information or experience with patients having papiledema after vaccination, and specifically which vaccine?
   First of all, vaccinations don't cause papilledema. Papilledema is optic disc swelling due to increased intracranial pressure. If you mean optic disc swelling after vaccination (presumed optic neuritis), I have only personally seen this 2 times in 30 years of practicing neuro-ophthalmology. It must be rather rare. There are multiple case reports in th eliterature from a wide variety of

vaccinations. This article would be illustrative: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5858858/