

WWW 11: Paediatric Neuro-ophthalmology

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Ramesh
Kekunnaya



Preeti
Patil-Chhablani



Dan
Milea



Akshay
Badakere



William
Good



Eva
Gajdosova



Klara
Landau



Richard
Hertle



*Preeti Patil-Chhablani (PPC), Akshay Badakere (AB), Klara Landau (KL), Eva Gajdosova (EG),
Dan Milea (DM), William Good (WG), Ramesh Kekunnaya (RK)*

1. Do you see a role for OCTA in the FFUP of these patients?

PPC: OCTA is unlikely to contribute to follow up, currently there is no consensus about OCTA in papilledema, it has shown certain patterns in other forms for disc edema such as NAION

KL: No, at least not yet.

EG: don't have experience with OCT A, but combination of OCT disc and macula is helpful, OCT disc might not be reliable in large disc swelling but later when swelling is resolved can help to monitor RNFL loss, initially thickens is confounded with swelling, but monitoring of GCL is very helpful especially of visual field testing is not reliable or if there is functional overlay

DM: no proven role of OCTA in papilledema follow-up. More evidence in the acute phase, to differentiate it from NAION. In papilledema, there is typically less peripapillary drop out of vascular flow than in NAION

KR: Not really. OCTA might help differentiating papilledema from other causes of disc edema, no role in follow up of these cases.

2. What about exploring LSO tendon and tuck, if it is really lax?

KL: Would be a possibility if exocyclotorsion is present.

EG: On FDT L SO was not lax but will try force traction test as suggested by Ramesh

KR: Yes, that would be my option along with IO myectomy.

3. Was there extorsion on fundus in LE?

KL: Don't know.

EG: not obvious on initial examinations or during examination under anaesthesia, later child cooperation was limited, recently has improved, plan is to recheck it, on photos RE was slightly extorted or normal and left eye slightly intorted ???, therefore skew was mentioned, plan to repeat photos and fundoscopy in a hope he will be cooperative

4. In optic nerve diseases, we expect blue yellow colour defect. Which test can be performed on the patient with colour vision disorder?

KL: Colour perimetry is interesting in research, in daily practice black on white perimetry is being used which is fine in people with colour vision abnormalities.

DM: tritanopia is not the commonest colour vision deficiency in acquired optic neuropathies, except glaucoma

WG: Afferent pupil defect

5. Role of IV steroids?

PPC: IV steroids can be used in a small subset of cases with IIH – it is useful in those with the ‘fulminant’ form of the disease, specially to buy more time for a definite surgical intervention

AB: In Fulminant IIH there is a role of IV steroids, but should be given along with acetazolamide

KL: For the first patient with sight threatening papilledema? NONE. For the second patient with presumed ocular myasthenia? NONE. For the third patient with abnormal eye movements? NONE.

DM: there is no proven role of steroids in papilledema, except, possibly, in fulminant situations, while preparing a surgical intervention.

WG: none

KR: No role except in fulminant cases where IVMP can reduce the inflammation and further damage.

6. As you have an important sinus thrombosis, a stent couldn't be helpful?

PPC: stents usually help in stenosis, thrombosis due to a systemic etiology would require medical therapy

KL: Very dangerous!

DM: venous stenting in sinus thrombosis has not yet been evaluated with thorough trials in children

WG: Might be

KR: Possible, but not yet proven in adults and children.

7. Is there an indication for early ON decompression on the left?

PPC: One could definitely argue for ON decompression, unfortunately the patient's systemic condition did not allow any anesthesia / surgical intervention

AB: No, medical management can be a good first option

KL: Yes, Optic Nerve Sheath Fenestration would be correct in a less sick patient.

DM: I agree it should have been discussed, given the Frisen 4 - 5 severity and the significant visual loss (20/60). As a result, the end stage was optic atrophy on the left side. However, there are limitations, of course, due to the ability of the centre to perform ONSF, the general medical condition of the child, etc.

WG: Not early but maybe later

KR: I would go for early aggressive medical therapy. If medical therapy fails or not so effective, in this particular case if the systemic condition allows I would perform early ONSF.

8. If you'd prefer a more aggressive approach, when will be the point that you'd suggest the surgery to take place?

PPC: onset of optic neuropathy, not responding to treatment

KL: The appearance of the fundus (Frisén grade 5) was alarming, as was the visual field loss. Visual acuity may have been reduced due to macular exudate.

DM: I think that Frisen 4-5 and the severe left optic neuropathy were enough arguments to discuss this.

WG: After making sure the child is healthy—no more systemic clots

KR: No or minimal response to early medical therapy.

9. Is there a need for intracranial pressure measurement in venous sinus thrombosis, if the diagnosis is definite?

PPC: This is controversial, some neurologists argue that one can treat without LP as well, generally, in our protocol, we do plan LP in these cases, if the systemic condition allows. The CSF analysis also rule out the rare chance of meningitis that may cause a similar picture, especially in thrombosis secondary to infections.

AB: Yes

KL: Not really.

DM: in case of stenting, these measures are regularly performed, but considering the adopted treatment, there was no firm indication for measuring the ICP. Even LP with opening CSF pressure measurement is not systematically performed, given the obvious imaging.

WG: No

KR: Not really as MRV is evident.

10. Even if vision is threatened, is it safe to administer IV steroids in a patient with uncontrolled diabetes and DKA?

PPC: The systemic condition would take priority, a team discussion with Neurology, intensive care and endocrinology would be beneficial

AB: In uncontrolled Diabetes, always ideal to control blood sugars and then give steroids.

KL: No

DM: probably not

WG: No

KR: No

11. Is Tensilon or an alternative available anywhere in the world?

AB: Neostigmine is an alternative to tensilon, we use Neostigmine in India as tensilon is not readily available

KL: In Switzerland we still have Tensilon. The sleep test is as sensitive and you do not need a pharmacologic agent – just have the patient rest for 1-2 hours with eyes closed and check on him/her immediately when opening the eyes again.

DM: yes, for France and Singapore

WG: I don't know

KR: Not available in India, we use Neostigmine.

12. How often does the panel do Neostigmine test? and what is the sensitivity?

AB: In our centre we do it in all cases, the sensitivity and specificity varies literature quotes it as 83% and 97% respectively (Natarajan et al “accuracy of ice test in Myasthenic ptosis”, Neurology India journal, 2016)

KL: I have no experience with Neostigmine. According to the literature it is useful.

DM: rarely – but excellent sensitivity

WG: I don't know

13. Did the patient's blood work-up show eosinophilia?

AB: No

KL: Not my patient. But I remember that the answer was no

WG: I don't know

14. Was there any pupil involvement?

AB: No

KL: Not my patient. But I remember that the answer was no

DM: agree that pupil involvement would have immediately ruled out myasthenia. Genuine integrity of pupillary function is exceptionally rare, if ever possible in such an extensive brainstem involvement as in this case.

WG: I don't know

15. Can Anthelmintics be given after 3 days of steroid cover?

AB: Not in lesions in and around the brainstem, it is contraindicated

KL: Has to be discussed with internal medicine/infectious disease experts.

DM: I would have started both simultaneously.

WG: I don't know

KR: Not in cerebral lesions. Anthelmintics are dangerous.

16. Could the panellists discuss the bony orbit dissimilarity seen in the MRI images?

KL: Well even looking at the boy's photo there is mild hypoplasia of the right half of the face and a hypoglobus on the right.

EG: there is possible familial anatomical physiological variation, no pathology was described, but plan is to repeat interval scan

WG: I didn't see it

KR: I saw hypoplasia over the right side

17. What about MED (monocular Elevation Deficit) in the Right Eye?

KL: Is an option for some of the signs in this boy. The forced duction test, performed by an experienced strabismus surgeon, was negative – thus the less common paretic type of MED can be assumed.

EG: MED was considered as differential diagnosis, but intermittently his RE elevations on duction are full, I consider that anatomical displacement of the right orbit/eye inferiorly might play a role, he is trying to fixate with right eye harder to keep it in primary position or on up gaze and maybe left eye elevators according to Hering's law then are causing hypertropia and then asymmetry of SO muscle picked up on MRI might play role as well on top of possible L DVD with IO OA.

WG: Yes

KR: One of the differential diagnosis. FDT ruled it out I guess.

18. What about anisocoria? How is it explained?

KL: Physiologic anisocoria (at least in the photos anisocoria is minimal)

EG: I believe it is physiological, he was very difficult to examine, very wriggly, at first presentations it was checked by orthoptists and I have seen him with dilated pupils. Later when I have assessed pupils myself I have confirmed it and is consistent R<L, in bright/dim light, but normal reactions, but keeping my mind open in case it is related to possible CCDD.

WG: physiological

KR: I didn't see much. May be within physiological limits.

19. What about systemic work up?

EG: as for case 3 – there were no health concerns, the patient was asymptomatic otherwise, so no systemic work up was performed. Panellist suggested to rule out MG.

WG: Maybe for myasthenia

20. Did the presenter check versions?

EG: case 3 – versions were presented on photo of 9 positions of gaze during presentation, I always check versions first and then ductions, ductions were described as variable, as for right eye elevation, but recently they look full on either eye, that is confusing. Maybe anatomical position of right orbit and eye might play a role in his clinical picture.

WG: Don't know

21. Could this be latent nystagmus in the right eye?

EG: latent nystagmus, manifest latent, was considered as option, it is a part of FMNS (fusion maldevelopment nystagmus syndrome) which was discussed, nystagmus recording was suggested, he had intermittent variable left eye nystagmus -rotatory as well as jerk, during binocular viewing beating towards fixing right eye, left beating under monocular fixation with left eye, which might fit with FMNS. When it was first time noticed, because it was only in right eye, optic pathway glioma needed to be rule out and spasmus nutans was considered shortly, but there was no head bobbing, only AHP and dysconjugate nystagmus and saccades.

WG: yes

22. Did the presenter check the head tilt with each eye covered?

EG: not possible in the clinic, till now, he is not tolerating occlusion in the clinic, without being upset, but good point, I will try when his cooperation will improve or ask parents to record another video at home with occlusion.

WG: Not my patient

23. Is it possible for this case to be that of a brainstem stroke and not myasthenia?

KL: The definitive diagnosis was brain stem neurocysticercosis.

DM: genuine absence of pupillary involvement in such an extensive brainstem disorder is exceptionally rare, or even possible if correctly explored.

WG: Very unlikely

24. Nuclear 3rd without pupil involvement? I do not think response to steroid confirms that the mass was the cause of the condition.

AB: No, possible that since steroids were started early, pupil involvement did not occur.

KL: WRT 'Nuclear 3rd without pupil involvement?' - Very very unusual. WRT 'Think response to steroid does not confirm the mass was the cause.' - Disagree. In many brain lesions with surrounding edema steroids may lead to an impressive temporary improvement.

DM: I agree that nuclear 3rd without pupillary involvement is exceptionally rare or even possible if the patient is thoroughly examined.

WG: I don't know

25. Did the presenter look at fundus torsion? Is there LE Extorsion?

KL: Good question, probably yes, LSO was hypoplastic on MRI.

EG: not obvious on initial examinations or during examination under anaesthesia, later child cooperation was limited, recently has improved, plan is to recheck it, on photos RE was slightly extorted or normal and left eye slightly intorted ???, therefore differential diagnosis has mentioned skew deviation, plan to repeat photos and fundoscopy

WG: Not my patient - don't know

26. What about role of genetic testing for infantile myasthenia?

AB: Diagnosis can be established clinically

WG: yes

27. Do you routinely get a CT chest? Any concerns about radiation exposure?

KL: Young patients with myasthenia should be checked for thymoma. Could / should be performed using MRI in children.

DM: chest imaging is not a diagnostic tool for myasthenia, but rather a systematic search for possibly associated thymoma or other conditions of the thymus

WG: Not my patient

KR: MRI can be performed instead of HRCT.

28. Did anyone check for hyporeflexia or ataxia as can be seen in Miller Fisher?

AB: No we didn't look for it

KL: Don't know, not my patient. But a general neurologic examination has to be performed in any patient with myasthenic symptoms.

DM: absolutely agree, this has not been mentioned. Search for antibodies GQ1b, antiGM1, etc. could be useful, although, again pupil involvement is quite typical in MFS

WG: Not my patient

29. What is the role of confrontational perimetry in paediatric neuro-ophthalmology?

AB: Yes, if the child is 1s enough and understands instructions, definitely.

KL: In smaller children it should be done, looking for saccades, unfortunately it is rarely performed and homonymous hemianopias or other severe VF defects are being missed.

EG: I am using it if child is young or not able to perform other type of perimetry

WG: helpful

30. Dr. Preeti: What was the cause of DM? and were there any other thrombotic risk factors? Possibly multiple episodes of ketoacidosis led to dehydration and precipitated CSVT?

PPC: The patient had Type 1 DM, no associated genetic syndrome. The thrombosis was presumed to be caused by dehydration and diabetic ketoacidosis. The complete prothrombotic workup to look for other causes was negative.

DM: agree that extensive thrombosis is not common in a child with DM, requiring extensive investigations. There was a history of pancreatitis (?), but not clearly detailed or further reinvestigated. Pancreatitis can be associated with thrombogenic factors of various origins.

31. Definitely agree with Dr. Good that holding anticoagulation for surgical intervention could lead to death due to progression of thrombosis. Diamox lowers both ICP and IOP, so unclear how much it affects pressure gradient / perfusion.

KL: WRT 'Definitely agree with Dr. Good that holding anticoagulation for surgical intervention could lead to death due to progression of thrombosis.' - Agree. WRT 'Diamox lowers both ICP and IOP so unclear how much it affects pressure gradient / perfusion.' - Agree.

DM: there are various ways to perform surgeries in patients on anticoagulation, this option is worth a multidisciplinary discussion.

32. The papilledema was a high grade; ONSF should have been done!

PPC: Unfortunately, the systemic condition did not allow it

KL: It was important to stabilize the patient's general condition first. But I agree that in a less dangerous situation optic nerve sheath fenestration was indicated at this point.

DM: fully agree, it's at least worth a discussion if orbital surgeons are available.

33. Was it an option to give IV hydration with oral Diamox?

PPC: That is what was done – in patient management with IV hydration, electrolyte management with Diamox

KL: Diamox was given in a slowly increasing dosage and under stationary observation of electrolytes and hydration.

DM: Acetazolamide has no firm indication in high intracranial pressure due to venous sinus thrombosis in children.

WG: Don't know

34. What is the prognoses of the child with optic neuritis? since I had ever treated a recurrent case.

PPC: Depends on the etiology of the neuritis, other associated factors.

KL: Cannot be answered in this general way without further details.

WG: Usually good but this isn't optic neuritis