5.8.3. Diplopia (III): Nuclear & Internuclear Causes

Internuclear Causes of Diplopia

medial longitudinal fasciculus (MLF)

connects the sixth nerve nucleus on one side of the pons to the medial rectus subnucleus on the contralateral side of the midbrain

- slow adducting saccadic velocity in 1 eye
  - ± horizontal diplopia
  - ± adduction limitation
  - ± convergence limitation
  - difficulty tracking fast-moving objects
- lesion of right MLF causes right INO
- INO is named for the side of limited adduction
- abducting nystagmus of the fellow eye
- ± skew deviation
  - hyperdeviation ipsilateral to the lesion
  - ± vertical-oblique diplopia
- skew deviation without INO: hyperdeviation contralateral to lesion

internuclear Ophthalmoplegia (INO)

bilateral INO

- bilateral adduction lag

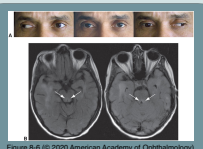

Figure 8-6 (© 2020 American Academy of Ophthalmology)

- bilateral abducting nystagmus
- vertical, gaze-evoked nystagmus
  - disruption of vertical vestibular pursuit and gaze-holding pathways
  - ascend from the vestibular nuclei through the MLF
- large-angle exodeviation
  - "wall-eyed" bilateral INO, or WEBINO, syndrome
  - midbrain lesion near the third nerve nuclei

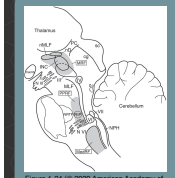

Figure 1-24 (© 2020 American Academy of Ophthalmology)

- 2 most common causes
  - adolescents and younger adults
    - demyelination
  - older adults
    - stroke
  - etiology
- pseudo-INO
  - myasthenic eyelid signs
  - no vertical gaze–evoked nystagmus
  - adduction paresis may resolve transiently after intravenous edrophonium (edrophonium test)
  - responds to appropriate systemic therapy
  - myasthenia gravis
  - differential diagnosis
- references

One-and-a-Half Syndrome

- pontine abnormality large enough to involve ipsilateral
  - ipsilateral INO
    - MLF
  - ipsilateral horizontal gaze palsy
    - PPRF (or sixth nerve nucleus)
  - only horizontal eye movement remaining is abduction of the contralateral eye

Figure 8-7 (© 2020 American Academy of Ophthalmology)

- WEBINO is midbrain
- vertical gaze is preserved
- one-and-a-half syndrome + involvement of intra-axial portion of the facial nerve
  - eight-and-a-half syndrome (7 + 1.5 = 8.5)
  - stroke
  - etiology

Nuclear Causes of Diplopia

third cranial nerve nuclear complex

- anatomy
  - subnuclei for 4 extraocular muscles
    - superior rectus fascicles decussate just after emerging from their subnuclei
    - nuclear lesions may bilaterally affect the superior rectus muscles
  - single subnucleus for the levator palpebrae muscles
    - nuclear lesions affect or spare both upper eyelids
    - central caudal nucleus
  - paired subnuclei for the pupillary constrictor muscles
    - Edinger-Westphal nuclei

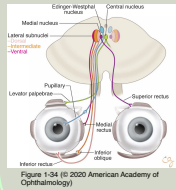

Figure 1-34 (© 2020 American Academy of Ophthalmology)

- lesions
  - uncommon
  - reduced perfusion through a small, paramedian-penetrating blood vessel
    - affects the oculomotor nerve nucleus & fascicle on one side
    - asymmetric

fourth cranial nerve nucleus

- short course within the brainstem
  - intraparenchymal lesions of the fourth nerve are rare!
- decussates in anterior medullary velum
- etiology
  - microvascular
  - inflammatory
  - demyelinating
- nuclear lesion is clinically identical to fascicular lesion
- descending sympathetic pathway is close to the caudal portion of the nucleus
  - fourth nerve palsy + contralateral Horner syndrome

sixth cranial nerve nucleus

- 2 populations of neurons
  - primary motoneurons
    - innervate the ipsilateral lateral rectus muscle
  - internuclear motoneurons
    - innervate the contralateral medial rectus subnucleus (via contralateral MLF)
  - ipsilateral horizontal gaze palsy
    - not an isolated abduction paresis!
    - patients may not experience diplopia!
- surrounded by the looping fibers (genu) of CN VII
  - ipsilateral upper and lower facial weakness
  - facial colliculus syndrome

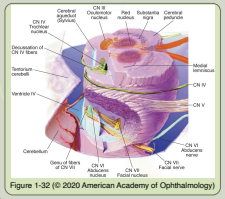

Figure 1-32 (© 2020 American Academy of Ophthalmology)

Ophthalmology Mind Maps/Arman Mashayekhi, MD