5.8.7. Diplopia (VII): Infranuclear Causes of Diplopia (IV)

Carotid cavernous sinus fistula

- abnormal connections between the carotid artery or its branches and the cavernous sinus
  - **direct, high-flow connections**
    - between the internal carotid artery and the cavernous sinus
    - after severe head trauma
  - **indirect, "dural" low-flow connections**
    - between small arterial feeders off the internal or external carotids and the cavernous sinus
    - occur spontaneously
    - older women
  - high- and low-flow fistulas cannot be distinguished reliably through physical examination
    - except cranial bruit that indicates high flow
  - cerebral angiography is necessary to make the correct diagnosis

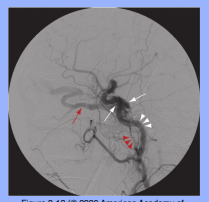

Figure 8-13 (© 2020 American Academy of Ophthalmology)

- clinical presentation
  - pain
    - may partly result from ocular surface drying if proptosis is significant
  - ocular motor neuropathy with diplopia
    - compression of CN VI by an enlarged inferior petrosal sinus
      - isolated sixth nerve palsy even in the context of a quiet eye
      - drains the fistula
  - may reverse blood flow within the superior ophthalmic vein
    - arterialization of conjunctival vessels
      - proptosis
      - venous congestion within the orbit

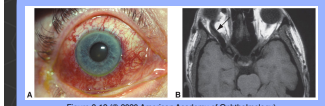

Figure 8-12 (© 2020 American Academy of Ophthalmology)

  - elevated intraocular pressure
  - arterial or venous compromise to the retina and eye
  - ischemic optic neuropathy
  - choroidal effusions
    - can push the iris forward and produce angle-closure glaucoma
  - cerebral venous infarction (rare)
- management
  - angiographic studies are required
  - some indirect fistulas remain stable or close spontaneously
  - interventional radiologic techniques
    - variety of thrombogenic materials
      - coils, beads, or balloons
  - radiosurgery

Neuromyotonia

- rare
- prior skull-base radiation therapy
  - months-years postradiation
- may affect any of the ocular motor nerves or their divisions
- episodic diplopia
  - often triggered by activation of the affected nerve
  - overaction of the nerve produces ocular misalignment
  - 30–60 seconds
- medical therapy
  - carbamazepine and its derivatives
    - first-line treatment

Paresis of More Than One Cranial Nerve

- benign microvascular disease rarely causes simultaneous involvement of >1 ocular motor nerve
- simultaneous involvement of contiguous nerves (CNs III, IV, V, VI, and sympathetic nerves)
  - cavernous sinus lesion
- etiology
  - bilateral cranial nerve involvement
    - diffuse infiltrative process
      - carcinoma, leukemia, or lymphoma
    - midline mass lesion that extends bilaterally
      - chordoma, chondrosarcoma, or nasopharyngeal carcinoma
    - meningeal-based process
    - inflammatory polyneuropathy
      - Guillain-Barré syndrome
      - sarcoidosis
    - myasthenia gravis
- diagnostic workup
  - neurologic evaluation should be undertaken
  - if neuroimaging results are normal
    - lumbar puncture with cytopathologic examination
    - special testing for cancer-associated protein markers
    - CT-PET scan
      - in suspected neoplastic meningeal involvement (meningeal carcinomatosis)
        - study of choice
    - repeat studies may be needed
- idiopathic multiple cranial neuropathy syndrome
  - diagnosis of exclusion

Tolosa-Hunt syndrome

- idiopathic, sterile inflammation that primarily affects the cavernous sinus
- severe, "boring" pain
  - almost always present
  - typically responds rapidly and dramatically to corticosteroid therapy
    - a positive response may also occur with neoplastic mass lesions, especially lymphoma
      - diagnosis of this syndrome is one of exclusion
- neuroimaging
  - enhancing mass within the cavernous sinus
- other cavernous sinus lesions
  - aneurysm
  - meningioma
  - lymphoma
  - schwannoma
  - pituitary adenoma
  - carotid cavernous fistula
  - metastasis
  - sarcoidosis
  - cavernous sinus thrombosis

Cavernous Sinus and Superior Orbital Fissure Involvement

- combination of third, fourth, and sixth cranial nerve palsies
  - If only 1 ocular motor nerve is involved, it is usually the sixth nerve
    - the only ocular motor nerve not protected within the dural wall of the cavernous sinus
- fifth cranial nerve involvement
  - facial hypoesthesia
- sympathetic involvement
  - third-order (postganglionic) Horner syndrome
- aggressive lesions of the cavernous sinus
  - especially infectious or inflammatory
  - may compromise venous outflow
    - engorgement of ocular surface vessels
    - orbital venous congestion
    - increased intraocular pressure
    - increased ocular pulse pressure
- clinically impossible to distinguish cavernous sinus lesions from superior orbital fissure lesions
  - sphenocavernous syndrome/parasellar syndrome
- offending lesion may extend toward the optic foramen or into the orbital apex
  - optic nerve dysfunction
  - orbital apex syndrome

Neuromuscular Junction Causes of Diplopia (Myasthenia Gravis)

- diplopia
  - any pattern of ocular misalignment
  - ptosis
- it never produces
  - sensory symptoms
  - pain
  - autonomic dysfunction
  - clinical pupillary dysfunction

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