



Chapter 105 – Brain and Cranial Nerve Disorders

Episode Overview

1. List the name, function and pathologic features of the cranial nerves
2. List 6 differential diagnosis for facial pain / trigeminal neuralgia
3. Trigeminal neuralgia - describe its diagnosis and management
4. Facial nerve paralysis: List 6 differential diagnoses for facial (CN VII) paralysis
5. Describe the facial weakness deficit associated with Bell's Palsy and list 3 other associated symptoms
6. List 4 components of treatment of Bell's Palsy, and list the symptoms that would make you suspect Lyme Disease
7. Differentiate between herpes zoster ophthalmicus and herpes zoster oticus
8. Describe the symptoms of vestibular schwannoma and diagnostic test of choice
9. What is the pathophysiology of diabetic mononeuropathy, and which nerves are usually involved?
10. What is Uhthoff's phenomenon?
11. List 10 clinical symptoms of Multiple Sclerosis and describe its diagnosis. Describe the typical CSF findings in a patient with MS.
12. Describe the management of an acute MS exacerbation and the usual management of a patient with relapsing-remitting MS who presents with a flare

Wisecracks:

1. Cerebral Venous Thrombosis Bounceback: What are the key pearls?
 - a. List four risk factors for cerebral venous thrombosis.
 - b. What CT findings may be seen in cerebral venous thrombosis? What is the most common CT finding?
2. Which cranial nerve is most commonly injured in trauma?
3. What syndrome is associated with bilateral acoustic neuromas?
4. How is diabetic mononeuropathy treated?

Rosen's in Perspective

What are three things I can guarantee you feel a little queasy when pimpled about?
Well we've got you covered here for Cranial Nerve problems, Cerebral Venous Thrombosis and Multiple Sclerosis.

When it comes to Cranial nerve deficits, remember to NOT MISS THE INTRACRANIAL CATASTROPHE! When deficits cannot be clearly attributed to lower motor neuron dysfunction, obtain definitive imaging (typically, a CT head +/- contrast or MRI).



Why do we give anti-virals to treat viral neuropathy?

Answer: From UptoDate

- Lessen the severity and duration of pain associated with acute neuritis
- Promote more rapid healing of skin lesions
- Prevent new lesion formation
- Decrease viral shedding to reduce the risk of transmission
- Prevent PHN

[1] List the name, function and pathologic features of the cranial nerves

Refer to figure 95.1 in Rosen’s 9th Edition for a more comprehensive table summarizing cranial nerve function and pathologic features when injured

Cranial Nerve	Function of Nerve	Presentation
CN I: Olfactory N.	Gustatory sense	Unilateral anosmia
CN II: Optic N.	Sight	Unilateral vision loss
CN III: Oculomotor N.	Motor function of levator palpebrae, superior rectus, medial rectus, inferior rectus, and inferior oblique muscles; pupillary constriction	Ptosis, eye deviated laterally and downward, diplopia, dilated and non-reactive pupil, loss of accommodation
CN IV: Trochlear N.	Motor function of superior oblique	Inability to move eye downward and laterally, diplopia, head tilt toward unaffected eye
CN V: Trigeminal N.	Sensation of face, scalp, and oral cavity; motor function of masseter and temporalis muscles/tensor tympani	Partial facial anesthesia, periodic lancinating facial pain
CN VI: Abducens	Motor function of lateral rectus muscle	Diplopia, inability to move affected eye laterally
CN VII: Facial N.	Motor function of muscles of facial expression; PSNS innervation of lacrimal and salivary glands; sensation of ear canal and tympanic membrane	Hemifacial paresis, abnormal taste, sensory deficit around ear, sensitivity to loud sounds
CN VIII: Vestibular N.	Hearing and balance	Vertigo, tinnitus, unilateral hearing loss
CN IX: Glossopharyngeal N.	Taste and sensation of posterior aspect of tongue, motor function of stylopharyngeus	(RARE) Intermittent pain in the pharynx
CN X: Vagus N.	Motor function of smooth and striated muscles and glands of the pharynx, larynx, and veli palatini; motor function of smooth muscle of the thoracic and abdominal	Loss of palate rise, dysphagia, hoarse voice



	viscera; sensory input from the pharynx, larynx, esophagus, and thoracic/abdominal viscera	
CN XI: Spinal Accessory N.	Motor function of the sternocleidomastoids and trapezius muscles	Shoulder drop and downward displacement of scapula
CN XII: Hypoglossal N.	Motor function of tongue	Tongue deviations

[2] List 6 differential diagnoses for facial pain / trigeminal neuralgia

Here is the list from Rosen's:

- Odontogenic infections
- Sinus disease
- Otitis media
- Acute glaucoma
- Temporomandibular joint disease
- Herpes zoster

Note: However, you need to remember the basics here. Craniofacial pain can come from sensory fibres from any of the following nerves. So, any disease entity involving any combination of the listed nerves needs to be considered:

- Trigeminal (CN V)
- Nervus intermedius (CN VII)
- Glossopharyngeal (CN IX)
- Vagus (CN X)
- Upper cervical spinal cord roots

[3] Trigeminal neuralgia - describe the diagnosis and management

Note: Trigeminal neuralgia = tic douloureux

Symptomatology: idiopathic pain in one or more distributions of the trigeminal nerve (CN V). The vast majority of cases (90%) result from vascular compression of the trigeminal nerve root

RF:

- Female-to-male ratio of 2:1
- 50 - 69 years old
- Right side of the face > Left side

Diagnosis: clinical syndrome

- Normal head and neck examination
- No neurologic deficits
- Episodic, unilateral facial pain associated with non-painful triggers



Management:

Mainstay = Carbamazapine 100mg PO BID.

Other options:

- Phenytoin
- Baclofen
- Valproate sodium
- Lamotrigine
- Gabapentin
- Levetiracetam

Disposition: Neurology with +/- Neurosurgery or ENT consult if surgical decompression is required

[4] Facial nerve paralysis: List 6 differential diagnoses for facial (CN VII) paralysis

- Bell's palsy
- Ramsey Hunt syndrome (herpes zoster oticus)
- Lyme disease (neuroborreliosis)
- Bacterial infections of the middle ear, mastoid, or external auditory canal
- Guillain-Barré syndrome
- HIV infection
- Tumor compressing nerve (from temporal bone, internal acoustic canal, cerebellopontine angle, or parotid gland)
- Stroke (**rare**) (the rare pons stroke can mimic LMN facial nerve palsy)
- Sjögren's syndrome (rare)**
- Sarcoidosis (rare)**
- Melkersson-Rosenthal syndrome (rare):** "condition with a female predominance characterized by recurrent episodes of facial paralysis, episodic facial swelling, and a fissured tongue" - UptoDate

[5] Describe the facial weakness deficit associated with Bell's Palsy and list 3 other associated symptoms

- Sudden onset (think hours) unilateral facial paralysis
- Eyebrow sagging
- Inability to close the eye
- Disappearance of the nasolabial fold
- Drooping at the affected corner of the mouth, which is drawn to the unaffected side
- Decreased tearing
- Hyperacusis
- Loss of taste sensation on the anterior two-thirds of the tongue



Note: There is a way to classify the nerve dysfunction

House-Brackmann classification of facial nerve dysfunction	
Grade	Finding
I	Normal
II	Mild dysfunction
III	Moderate dysfunction
IV	Moderately severe dysfunction
V	Severe dysfunction
VI	Total paralysis

[6] List 4 components of treatment of Bell's Palsy, and list the symptoms that would make you suspect Lyme Disease

- Glucocorticoids
- Antivirals for severe disease (controversial with poor evidence but recommended for Grade IV and higher)
- Eye care (artificial tears)
- Physiotherapy (controversial)

See Table 95.2 in Rosen's 9th Edition for dosing of steroids and antivirals in Bell's Palsy

Consider Lyme (or infectious mononucleosis) disease if:

- Systemic symptoms
- Bilateral facial paresis

Remember: caused by the spirochete *Borrelia burgdorferi* from Ixodes genus ticks

[7] Differentiate between herpes zoster ophthalmicus and herpes zoster oticus

Herpes zoster ophthalmicus = shingles of the trigeminal nerve

- Hutchinson's sign: cutaneous involvement of nasociliary nerve
- Look for ocular involvement = punctate epithelial erosions and pseudodendrites

Herpes zoster oticus = Ramsay Hunt syndrome

- Unilateral facial paralysis
 - Can precede rash by days
- Herpetiform vesicular eruption



- Pinna, external auditory canal, tympanic membrane, soft palate, oral cavity, face, and neck as far down as the shoulder
- Vestibulocochlear dysfunction

[8] Describe the symptoms and diagnostic test for vestibular schwannoma

Note: vestibular schwannoma = acoustic neuroma

Symptomatology: patients with sudden sensorineural hearing loss

Diagnosis: ENT referral and MRI with gadolinium

[9] What is the pathophysiology of diabetic mononeuropathy and which nerves are usually involved?

Note: Diabetics are at high risk of intracranial badness!!! Look for brainstem pathology

Diagnosis: DIAGNOSIS OF EXCLUSION

Symptomatology: Think about in diabetics with oculomotor nerve (CN III) palsy with sparing of the pupillary response

[10] What is Uhthoff's phenomenon?

Answer: *** Elevated body temp = worsening demyelinating disease symptoms ***

Definition: "Uhthoff's phenomenon (also known as Uhthoff's syndrome, Uhthoff's sign, and Uhthoff's symptom) is the worsening of neurologic symptoms in multiple sclerosis (MS) and other neurological, demyelinating conditions when the body gets overheated from hot weather, exercise, fever, or saunas and hot tubs. It is possibly due to the effect of increased temperature on nerve conduction. With an increased body temperature, nerve impulses are either blocked or slowed down in a damaged nerve but once the body temperature is normalized, signs and symptoms may disappear or improve."

[10] List 10 clinical symptoms of MS, describe its diagnosis, and describe the typical CSF findings in a patient with MS

Note: Multiple sclerosis should be suspected in patients who present with episodes of neurologic dysfunction that evolve over days and resolve over weeks. Apparent exacerbations of known MS can be brought on by other medical problems, most commonly infections.

Diagnosis: The diagnosis of MS is accomplished using the McDonald Criteria. In summary, the patient must demonstrate disseminated central nervous system lesions over both space



and time. This can be done based on either clinical findings alone or a combination of radiologic findings (use MRI) and clinical evidence of disease.

Symptomatology:

- Symptoms that suggest MS:
 - Relapsing and remitting symptoms
 - Onset between 15-50 years of age
 - Optic neuritis
 - Lhermitte's sign (electric shock sensations that radiate down back/limbs with flexion of the neck)
 - Intranuclear ophthalmoplegia
 - Fatigue
 - Uhthoff's phenomenon
- Typical MS Symptoms:
 - Sensory disturbances in limbs
 - Vision loss
 - Motor disturbances
 - Diplopia
 - Gait disturbance
 - Imbalance
 - Sensory disturbances in face
 - Lhermitte's sign
 - Vertigo
 - Bladder issues
 - Limb ataxia
 - Acute transverse myelopathy
 - Pain

CSF:

- CSF analysis is abnormal in 90% of the cases
- 50% patients have pleocytosis
- More than 5 lymphocytes /HPF
- ~70% of patients have increased immunoglobulin G (IgG) ranging from 10% to 30%
- Increased oligoclonal bands of IgG in 85% to 95% (oligo bands are also seen in neurosyphilis, fungal meningitis, and other CNS infections)

[12] What is the management of an acute MS exacerbation and what is the usual management of a patient with relapsing-remitting MS who presents with a flare?

Answer:

- Consult their primary neurologist!
- IV methylprednisolone, 250 to 500 mg every 12 hours for 3 to 7 days effectively promotes earlier resolution of recurrences.
- IV methylprednisolone has been shown to speed the recovery from vision loss from optic neuritis associated with MS.



Relapsing remitting:

- Initiate treatment with interferon- β or glatiramer acetate.
- Newer disease-modifying agents include a bunch of -MABs: laquinimod, daclizumab, natalizumab, and terifunomide
- Facial pain: Carbamazepine
- Tremor: Propranolol or Benzos
- Spasticity: Baclofen

Wise Cracks:

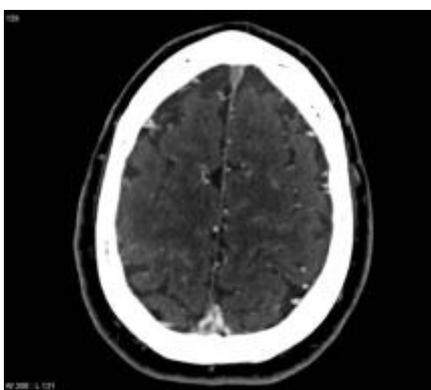
[1] Cerebral Venous Thrombosis Bounceback: What are the key pearls?

Four risk factors for cerebral venous thrombosis:

- Transient risk factors
 - Infection - think any HEENT or systemic infection
 - Pregnancy
 - Dehydration
 - Head injury / trauma / post-neurosurgery
 - Drugs:
 - OCPs, HRT, androgens, tamoxifen, glucocorticoids
- Permanent risk factors
 - Inflammatory disease: SLE, Behçet's disease, GPanG, IBD, sarcoidosis
 - Malignancy
 - Hematologic conditions
 - Procoagulant disease
 - Antithrombin deficiency
 - Protein C/S deficiency
 - Factor V Leiden mutation
 - Hyperhomocysteinemia
 - Polycythemia
 - Congenital heart disease
 - Thyroid disease

What CT findings may be seen in cerebral venous thrombosis? What is the most common CT finding?

- Delta Sign / Dense Triangle / Empty Triangle Sign Ref: [here](#)





- Cerebral Edema



- Absent Venous Flow
- Hemorrhage secondary to venous congestion Ref: [here](#)



Pearls:

- ***Non-contrast-enhanced CT scanning is not adequate to rule out CVT*****
- *** MRI with MRV is recommended*****
- Treatment = systemic anticoagulation, even in the setting of hemorrhagic cerebral infarcts, unless another contraindication exists!!!
- Hemorrhage secondary to venous congestion Ref: [here](#)

[2] Which CN is most commonly injured in trauma?

Answer: *** Trigeminal Nerve: anesthesia dolorosa ***

Definition: According to UptoDate: "Painful post-traumatic trigeminal neuropathy (anesthesia dolorosa) — Painful post-traumatic trigeminal neuropathy, also known as anesthesia dolorosa, is characterized by unilateral facial or oral pain that occurs after traumatic injury of the trigeminal nerve, accompanied by additional symptoms or signs of trigeminal nerve dysfunction. This condition is a perfect example of how difficult central pain can be to describe since the patient has sensory loss, but at the same time, can feel pain. In



anesthesia dolorosa, pain is superimposed in an area of the face that either lacks or has impaired sensation.”

[3] What syndrome is associated with bilateral acoustic neuromas?

Answer: approximately 5% of cases and generally associated with Type 2 Neurofibromatosis

[4] How is diabetic mononeuropathy treated?

Answer: NO CURE. Improve glucose control. Resolution within 3-6months. Treatment consists of:

- Patching the affected eye
- Administration of analgesics
- Antiplatelet therapy