PART 2

CN

- V
- VII
- V
- XII

Horner Syndrome
Neuromuscular Disease
Clinical presentations, signs and symptoms

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COI

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Is it neuromuscular?
Combinations?

Clinical presentation

Case history

new concepts

examples
Neuromuscular System
(A common construct)

- (Extracranial parts of the Cranial nerves)
- Nerve Roots
- Plexus
- Peripheral nerves
- Neuromuscular junction
- Muscle
- Autonomic system
Neuropathies: How often?

Median (CTS) 103.00
Ulnar 25.00
Radial 2.50
Cut. Femoral 10.00

Disc (> 50 yrs)
Cervical (aged) 600.00
Lumbar (aged) 1079.00

PNP DM II: 6400

Pro 100 000 Patient years
Source: Callaghan; Ann Arbor 2016, USA
Combinations of neuromuscular and central nervous system. Examples:

- MND/ALS
- Some hereditary neuropathies
- Myotonic dystrophy
- etc.
Case history

Family history (tree)

Diseases, medications,

Exposition, trauma

Onset, duration, acute, chronic progressive, fluctuating, episodic

Symptoms:
  motor, sensory, autonomic movement, composite

Acquired/genetic

Before examining the patient-create hypothesis
Time

Acquired

Acute

Hereditary

Chronic

episo
Symptoms and signs

Motor  Sensory  Composite function

Autonomic  Neuropathic Pain  Distribution
Symptoms and signs

Skin

Skeletal system
Motor

Distribution of weakness, atrophy

Strength (BMRC)

Time: Fluctuating, episodic

Muscle volume

Atrophy

Hypertrophy

Phenotype distribution: myopathy, polio, club foot
Winging
Proximal weakness
Carcinomatose Neuromyopathy

EMG
Percussion myotonia
Muscle hypertrophy

Examples

Pseudohypertrophy
Endocrinopathy
Focal muscle hypertrophy
Overuse infections
Amyloid
Steroid myopathy
Movement

- Fasciculation
- Myokymia
- Rippling
- „Stiffness“, myotonia
- Percussion myotonia
- Cramps

Tongue Ultrasound
Sensory

Quality

Sensory assessment

Instruments

Type of fiber lesion

Tinel Hofmann sign

Sclerotoma,
myotoma,
„neurotoma“
Instruments
A: superficial peroneal nerve

B. Infrapatellar nerve

Tinel Hofmann: red circle
Foot
Test Composite function!

Simple (but complex) functions:

- Opening and closing buttons
- Recognition of objects
- Balance: Romberg
- Stance on one leg
- Can you put on your trousers standing freely one leg?

Muscle:

- „Hands up“
- Standing up from squat
- Push ups
Ask for Autonomic dysfunction

- Dry mouth, dry eyes?
- Voiding
- Sexual dysfunction
- Orthostasis
- Gastrointestinal
Neuropathic Pain

Pain characteristics
(stabbing, shooting, electric)

Distribution- cave Allodynia

Sensory, motor involvement

Elicited by: eg Tinel Hofmann
Neuropathic Pain conditions

- Acute episodic
- Painful neuropathy
- Small fiber
- Trauma causalgia
- Localized
- Surgery
- Erythromelalgia
- Phantom- pain
- PRURITUS
Intercostobrachial nerve
Painful neuropathies

- Alcohol
- Metals
- Drugs (eg. CIPN)
- Immune-mediated, Vasculitis
- Hereditary (eg Fabry)
Skin

Haemangioma

Waterhouse F. syndrome
Thickening of nerves

Greater auricular nerve (intraoperative)
Radiofibrosis syndrome
Hyperkeratosis in denervation
Skin

Lymphoma
Hand Foot Syndrome
Skeletal system
Focal dysfunction/Weakness
Distribution: Neuropathies and Radiculopathies
Further Ancillary Tests

• NCV/EMG

• Imaging: ultrasound, MRI, CT

• Autonomic test

• Biopsy

• Laboratory

• Genetics
Neuromuscular lesions

- CN
- Plexus lesions
- Mononeuropathies
- Polyneuropathies
- Neuromuscular transmission
- Muscle
Facial nerve: 3 different problems
Cases of tongue involvement
The Plexus

• Cervical

• Brachial

• Lumbar

• Sacral

• (Coccygeal)
Phrenic nerve palsy
cervikalis
Local tumor affecting
the cervical plexus
Seroma compression of the brachial plexus
Lumbar, sacral - Anatomy
Sacral Plexus Vascular
Mononeuropathy
Acute Swelling and median nerve dysesthesia
Acute Swelling and median nerve dysesthesia.
Ultrasound: synovial cyst, yellow: median nerve.
Ulnar nerve lesion

Median nerve: Thenar atrophy
Chemotherapy paravasate, damage of cutaneous nerve
Radial nerve palsy after fracture
Ultrasound shows preserved continuity of the radial nerve
Diagnosis of nerve tumors

SCHWANNOM

[Image of an arm with a red line indicating the location of a tumor and an ultrasound scan of the tumor]
Hip operations

- Hip operations
- Femoral nerve
- Sciatic nerve (traction)
- Cutaneous nerve

From: Sunderland

Fig. 191
The branch fibre composition of the funiculi of the sciatic nerve and its two main divisions. For key to symbols see Figures 192 and 193.
Arrow points towards screw from the acetabulum in contact with the sciatic nerve
Nerve reconstruction

- Neuroplasticity:
- Brachial plexus surgery:
  - Phrenic nerve and intercostal
  - Contralateral C 7 transfer
- Hand transplant
M. Tibialis posterior transfer
Polyneuropathy

- Distribution
- Statistics
- symmetric
- Asymmetric
<table>
<thead>
<tr>
<th>Frequency</th>
<th>Diabetes</th>
<th>Toxic neuropathies</th>
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<tbody>
<tr>
<td>HIV</td>
<td>Immune mediated</td>
<td>Infection</td>
</tr>
</tbody>
</table>
Hereditary

Small fiber

Painful neuropathies

Neoplastic

Immune mediated

Paraneoplastic
Frequency of neuropathy

- Most frequent: Diabetes, cryptogenic and toxic.
- Inflammatory, genetic, etc less frequent.

Diabetes

- Symmetric
- Distal symmetric neuropathy
- Diabetic autonomic neuropathy
- “Pseudotabic“ type
- CIDP like
Diabetes

- Asymmetric forms
- Multiplex
- Truncal neuropathy
- Diabetic amyotrophy
Diabetes

• Prediabetic neuropathy

• „Insulin neuritis“
Diabetes

- CN: III, (VII)
- Mononeuropathy: CTS
- Diabetic muscle infarct
- Diabetic cachexia
Chemotherapy induced neuropathy

- Acute
  - Oxaliplatin
  - Taxane

- Cumulative
  - Platin
  - Taxane
  - Vinka alkaloid
  - Thalidomide
  - Bortezomibe

- "Coasting"

- Late effects
  - Sensory
  - Coordination
  - Pain
  - CN: taste, hearing
  - Erectile dysfunction
  - Reduction of LQ
Chemotherapy induced neuropathy

Different locations of pathology

Alpha-2-delta-Typ1 Subunit Taxane

Axon

Peripherer Nerv

Unbemerkte Fasern C Fasern

Mitochondrien Schwellung Taxane

NMT

Arbor

Langehans cell activation

SP: Spinalganglion
NMT: Neuromuskelärer Übergang
CIPN-PNP

- Acute (Oxaliplatinum)
- Chronic and cumulative
- Coasting
- Late CIPN, RT, Muscle
- Induced (PD)
Neoplastic neuropathy
Toxic neuropathies

• Alcohol

• Industrial agents

• Historical:
  • Spanish toxic oil syndrome
  • U boat oil TCE neuropathy
Infection

- HIV
- Leprosy
- CMV
- Herpes
  - VII
  - Mononeuropathy
- Motor involvement
- Myelopathy
HIV and the PNS

ALS-variant syndrome
Autonomic

CIDP
Cytomegalovirus
DILS
Drug-induced
Guillain-Barré
Mononeuritis multiplex
Neuromyotonia
Sensory motor polyneuropathy
VII nerve palsy
Mononeuropathy
Brachial plexopathy

Toxic Drug treatment
Hereditary neuropathies

- CMT
- Fabry
- Amyloid
- Porphyria
CMT detected in an asymptomatic patient.
Left: foot deformity.
NMT

- MG (AchR pos)
- MusK LRP4
- „Seronegative“
- LEMS
- Neuromyotonia
- Other NMT disorders
Ocular myasthenia
• Triple furrow tongue
Lambert Eaton myasthenic syndrome

• Autoimmune presynaptic disorder

• 50 % paraneoplastic (SCLC)

• Electrophysiology: Increment

• Clinical: proximal weakness, facilitation of reflexes
Neuromyotonia

Peripheral and CNS disorders with VGKC antibodies

- PNS
  Neuromyotonia or Cramp
  Fasciculation Syndrome
- Muscle twitching
- Cramps
- Stiffness
- Sweating
- Pain

- CNS
  Limbic syndromes
  - Memory loss
  - Seizures
  - Personality change
  - Psychosis
  - REMS sleep behaviour disorder

Morvan’s Syndrome
- Insomnia
- Constipation
- Arrhythmias
Muscle

• Distribution
• Onset/Course
• Acquired/genetic
Infection  Systemic disorders  Myotonia

Critical illness  Sarcopenia  Cachexia
Hyperlordosis              Shoulder girdle weakness                 Shortened achilles tendons
A quick test for myopathic weakness
Mitochondrial myopathy
Dermatomyositis
Myositis

- DM (25%) (Anti-TiF1γ)
- PM (10%)
- Necrotizing myopathy (NAM)
Volar atrophy
Axial myopathy
Toxic myopathies, Hyper CK emir

• Toxic myopathies: see: http://neuromuscular.wustl.edu

• Hyper CK emia: EFNS guidelines on the diagnostic approach to pauci- or asymptomatic hyperCKemia European Journal of Neurology 2010, 17: 767–773 (download)
Motor Neurone Disease

- MND
- ALS
  - Variations
  - Genetic forms
- Kennedy syndrom
• ALS

• Variants

• Genetic types
Flail arm syndrome

Atrophy of shoulder girdle (flail arm)

Thigh and leg muscles preserved
Sources:
Join the World Brain Day 2018!

Brain impacted by air pollution

Check the WFN website for information.