DISEASES OF PERIPHERAL NERVES

Numerous inherited and acquired pathological processes may affect the nerve roots (radiculopathy), the nerve plexuses (plexopathy) and/or the individual nerves (neuropathy).

Cranial nerves 3-12 share the same tissue characteristics as peripheral nerves elsewhere and are subject to the same range of diseases. Nerve fibres of different types (motor, sensory or autonomic) and of different sizes may be variably involved. Disorders may be primarily directed at the axon, the myelin sheath (Schwann cells) or the vasa nervorum

PATTERN OF INVOLVMENT

Mononeuropathy Simplex=Single Nerve

Mononeuropathy Multiplex=Several Nerves Randomly &Noncontiguously

Polyneuropathy(Peripheral Neuropathy)=Dysfunction of Numerous Peripheral Nerves at the Same Time leading to predominantly distal & symmetrical deficit usually affecting lower more than upper limbs

Types &Causes of peripheral neuropathy
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	focal	Multifocal	(mononeuropathy multiplex	Generalized	(polyneuropathy)
neurophysiology	mononeuritis	acute	chronic	acute	chronic
Demyelinating= slowed conduction , coduction block	entrapment	diphtheria	Leprosy Paraproteinaemia MMN HNPP	Guillain barrie suramine	Hereditary Lymphoma CIDP Myeloma Arsenic Amiodarone IgM paraproteinaemia diphtheria
Axonal = reduced or absent action potential normal ncv	Severe entrapment	Diabetes Vasculitis Lyme disease cryoglobulinaemia	Diabetes Neoplatic HIV Sarcoidosis amyloid	Alcohol G.B. Variants Toxins Critical inness Porphyria Paraneoplastic Tick paralysis	Metabolic Endocrine Alcohol Drugs Toxins Vit. Deficiency Hereditary igG paraproteinaemia Paraneoplastic amyloidosis

Diseases of Peripheral Nerves

SYMPTOMS &SIGNS

1.Sensory Disturbances
A.Numbness, Hyperpathia, Impaired Sensation & SPONTANEOUS PAIN esp. in SMALL FIBER involvement ;D. M. , Porphyria, AIDS, Alcoholic, Entrapment …..
B. Dissociated Sensory Loss Small Fib. Pain &Temp.
Large Fib. Touch, Vib. & Position
2. MOTOR DEFICITS
Weakness , Wasting , Fasciculation
Diminished or Absent Reflexes
i.e. LMNL
3.AUTONOMIC DISTURBANCES
Post. Hypotension, Coldness, Imp. Sweating, Impotence….esp. GBS, Diabetes, Renal Failure, Porphyria….
4. ENLARGED NERVES
Leprosy, Amyloidosis, HSMN, Refsum Dis., Acromegaly..

Causes of P. N .

1. Inflammatory: GBS , CIPD
2. Metabolic &Nutritional :D.M. ,Uremia, Liver Failure, Hypothyroidism, Acromegaly, B12 Deficiency….
3.Infectious &Granulomateous:AIDS, Leprosy, Diphtheria, Sarcoidosis…
4.Vasculitis: PAN, SLE, Rh.Arthritis…
5.Neoplastic &Paraneoplastic
6. Drugs &Toxins:Alcohol, INH, Vinicristine, Phenytoin, Heavy Metals….
7.Hereditory:HSMN, HSN, Refsum Disease,Porphyria..
8. IDIOPATHIC

Drugs causing peripheral neuropathy

Cardiovascular agents	Amiodarone , statins , hydralazine
Chemotheraputic agents	Cisplatine , thalidomide , vincristine
Anti-infective agents	Chloramphenicol , INH , ETB , nitrofurantoin , metronidazole
Others	Gold , disulfiram , pyridoxine , tacrolimus , colchicine , phenytoin

DIFFERENTIAL DIAGNOSIS

- Diseases of muscles &n.m. junction
- normal sensation& tendon reflexes
- Diseases of spinal cord
- pyramidal signs &sensory level below the lesion
- Radiculopathies
- dermatomal &myotomal distribution

INVESTIGATIONS

- CONFIRM DIAGNOSIS
- EMG = DENERVATION
- ENG &NCV = Demyelination or Axonal Degeneration
- REVEAL UNDERLYING CAUSE

TREATMENT

- UNDERLYING CAUSE
- NURSING CARE ? ULCERS &CONTRACTURES
- RESPIRATORY MONITORING &MANAGEMENT
- CARE OF SKIN &NAILS

- RELIEF OF PAIN ----Lancinating Pain--PHENYTOIN
- CARBAMAZEPINE
- MEXILETINE
- Constant Pain----AMITRIPTYLINE
- GABAPENTINE,PREGABALIN

GUILLIAN BARIE SYNDROME ACUTE ASCENDING POLYRADICLONEUROPATHY

This syndrome of acute paralysis develop in 70 % of patients 1-4 weeks after respiratory infection or diarrhoea (particularly Campylobacter).
In Europe and North America, acute inflammatory neuropathy is most commonly demyelinating (AIDP).
Axonal variants are more common in China and Japan .

Clinical features

Distal paraesthesia and limb pains precede a rapidly ascending muscle weakness from lower to upper limbs , more marked proximally than distally.
Facial and bulbar weakness commonly develops , and respiratory weakness requiring ventilatory support occurs in 20 % of cases .
In most patients weakness progresses for 1-3 weeks but rapid deterioration to respiratory failure can develop within hours

On examination there is diffuse weakness with widespread loss of reflexes.
Overall, 80% of patients recover completely within 3-6 months, 4% die, and the remainder suffer residual neurological disability which can be severe. Adverse prognostic features include older age, rapid deterioration to ventilation and evidence of axonal loss on EMG**.**

Investigations

The CSF protein is abnormal at some stage of the illness, but may be normal in the first 10 days. There is usually no rise in CSF cells (lymphocytosis of > 50/ml suggests an alternative diagnosis).
Electrophysiological studies are often normal in the early stages but show typical changes after a week or so , with conduction block and multifocal motor slowing , sometimes most evident proximally as delayed F-waves.

Management

During the phase of

deterioration , regular monitoring of respiratory function (vital capacity and arterial blood gases) is required, as respiratory failure may develop with little warning and require ventilatory support.

Ventilation may be needed if the vital capacity falls below 1L , but ventilation is more often required because of bulbar weakness leading to aspiration.

General management to protect the airway and prevent pressure sores and venous thrombosis is essential .

Corticosteroids have been shown to be ineffective .

Plasma exchange and intravenous immunoglobulin therapy shorten the duration of the illness ,reduce severity and improve prognosis provided treatment is started within 14 days of the onset of symptoms .

DIABETIC NEUROPATHY

- AMYOTROPHY = PAIN &WEAKNESS WITH ATROPHY PELVIC GIRDLE &THIGH MUSCLES WITH ABSENT KNEE REFLEX &LITTLE SENSORY LOSS
- MONONEUROPATHY = ACUTE PAINFUL CRANIAL NERVES
- BOTH HAVE GOOD PROGNOSIS

Entrapment neuropathy

Focal compression or entrapment is the usual cause of mononeuropathy . However , some patients present with what initially appears to be a single nerve lesion and then go on to develop multiple nerve lesions.This is termed mononeuritis multiplex .

Symptoms and signs

Nerve	Symptoms	Muscle weakness/muscle-wasting	Area of sensory loss
Median at wrist) (carpal) (tunnel syndrome	Pain and paraesthesia on palmar aspect of hands and fingers, waking the patient from sleep. Pain may extend to arm and shoulder	Abductor pollicis brevis	Lateral palm and thumb, index, middle and medial half 4th finger
Ulnar (at elbow)	Paraesthesia on medial border of hand, wasting and weakness of hand muscles	All small hand muscles, excluding abductor pollicis brevis	Medial palm and little finger, and medial half 4th finger

Nerve	Symptoms	Muscle weakness/muscle-wasting	Area of sensory loss
Radial	Weakness of extension of wrist and fingers, often precipitated by sleeping in abnormal posture, e.g. arm over back of chair	Wrist and finger extensors, supinator	Dorsum of thumb
Peroneal	Foot drop, trauma to head of fibula	Dorsiflexion and eversion of foot	Nil or dorsum of foot
Lateral cutaneous nerve of the meralgia) **thigh** (paraesthetica	Tingling and dysaesthesia on lateral border of the thigh	Nil	Lateral border of thigh

In an entrapment neuropathy, pressure initially damages the myelin sheath, and neurophysiology will show slowing of conduction over the relevant site. Sustained or severe pressure damages the integrity of the axons, demonstrable as loss of the sensory action potential distal to the site of compression.

Certain conditions increase the propensity to develop entrapment neuropathies. These include acromegaly, hypothyroidism, pregnancy, any pre-existing mild generalised axonal neuropathy (e.g. diabetes), and oseophytes.

Entrapment neuropathy

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- MEDIAN N. COMPRESSION AT THE WRIST
- IDIOPATHIC, PREGNANCY, TRAUMA, ARTHRITIS, MYXOEDEMA, ACROMEGALY, TENOSYNOVITIS……
- PAIN, NUMBNESS IN MEDIAN N. DISTIBUTION ?SHOULDER
- > AT NIGHT
- WEAKNESS & ATROPHY OF THENAR MUSCLES
- TINEL SIGN , PHALEN MANEUVER
- Rx LOCAL STEROIDS, WRIST SPLINT, SURGERY

Facial nerve palsy

Idiopathic facial nerve palsy or Bells palsy is a common condition affecting all ages and both sexes .The lesion is within the facial canal and may be due to reactivation of latent herpes simplex virus 1 infection .Symptoms usually develop subacutely over a few hours , with pain around the ear preceding the unilateral facial weakness. Patients often describe the face as numb but there is no objective sensory loss (except possibly to taste).

Hyperacusis can occur if the nerve to stapedius is involved , and there may be diminished salivation and tear secretion . Examination reveals an ipsilateral lower motor neuron facial nerve palsy . Vesicles in the ear or on the palate indicate that the facial palsyis due to herpes zoster rather than Bells palsy .

Prednisolone 40-60 mg daily for a week speeds recovery if started within 72 hours.Artificial tears and ointment prevent exposure keratitis and the eye should be taped shut overnight. About 80% of patients recover spontaneously within 12 weeks. A slow or poor recovery is predicted by complete paralysis, older age and reduced facial motor action potential amplitude after the first week. Recurrences can occur but should prompt further investigation. Aberrant re-innervation may occur during recovery, producing unwanted facial movements (e.g. eye closure when the mouth is moved) or 'crocodile tears' (tearing during salivation).