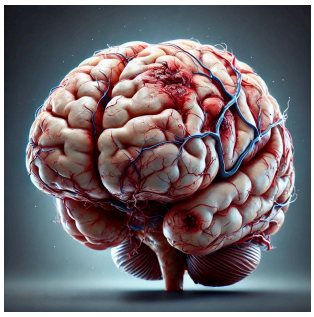

**CNS—pathology~4**
**Written by: Dr.Ali Abujammil.**

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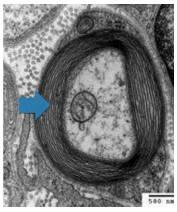
**Myelin is a protein-lipid complex that wraps around the axons of neurons. It acts as an insulating layer, allowing faster transmission of electrical signals in the nervous system.**

**Function of Myelin: Myelin insulates axons, preventing electrical signals from leaking out and It enables rapid signal propagation by allowing electrical impulses to "jump" between gaps in the myelin (called Nodes of Ranvier) instead of traveling continuously along the axon. This process is known as saltatory conduction, making signal transmission much faster.**

**Composition of Myelin: Myelin is made of multiple layers of plasma membrane, It is formed by two types of glial cells: Oligodendrocytes in the central nervous system (CNS) (brain and spinal cord) and Schwann cells in the peripheral nervous system (PNS) (nerves outside the brain and spinal cord).**

Myelin in this electron microscopic picture appears as layers of plasma membrane wrapped around the axon.



**Myelin in the PNS (Peripheral Nervous System): Schwann cells produce myelin by wrapping around the axon multiple times, Each Schwann cell forms a segment of myelin for a single axon.**

**Myelin in the CNS (Central Nervous System): oligodendrocytes produce myelin, Unlike Schwann cells, each oligodendrocyte can form myelin for multiple axons at the same time.**

**White Matter & Myelin: Myelinated axons make up most of the white matter in the brain and spinal cord, The high lipid content of myelin gives white matter its characteristic pale appearance.**

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**Diseases of Myelin in the PNS**

**What is Segmental Demyelination? is the main type of myelin injury in the peripheral nervous system (PNS) In this condition, the myelin sheath is damaged or lost, but the axon itself remains intact and can still survive Over time, Schwann cells may attempt to remyelinate the axon, but this process is often incomplete or inefficient.**

**Causes of Demyelinating Neuropathies-Demyelinating diseases in the PNS are mainly caused by:**

**1. Hereditary Causes (Genetic Disorders): Some genetic conditions lead to defective myelin formation or maintenance; Example: Charcot-Marie-Tooth disease (CMT) – a group of inherited disorders that affect peripheral nerves, causing weakness, numbness, and muscle wasting.**

**2. Immune-Mediated Destruction: The body's immune system mistakenly attacks the myelin sheath, leading to its breakdown; Example: Guillain-Barré Syndrome (GBS) – an**

**autoimmune disorder where the immune system rapidly destroys myelin in the PNS, leading to muscle weakness and even paralysis.**

**Consequences of PNS Demyelination: Slower nerve signal transmission, leading to weakness, numbness, and loss of reflexes and Partial recovery is possible if Schwann cells can regenerate new myelin but If severe or repeated damage occurs, axons may also degenerate, leading to permanent nerve dysfunction.**

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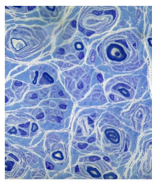
**Segmental Demyelination- What Happens in Segmental Demyelination?**

**When myelin is damaged in segmental demyelination, Schwann cells proliferate and attempt to remyelinate the axon If the damage is mild or moderate, function can be restored after remyelination However, repeated cycles of demyelination and remyelination can lead to structural abnormalities in the nerve.**

**Onion Bulb Appearance & Hypertrophic Neuropathy: In cases where demyelination-re-myelination happens repeatedly, more Schwann cells accumulate around the axon This results in thickened nerve fibers, which can be seen under the microscope as an onion bulb appearance (layers of Schwann cells surrounding the axon like onion layers) This condition is associated with hypertrophic neuropathy, where the nerves become enlarged due to excessive Schwann cell proliferation.**



**Clinical Features of Segmental Demyelination-Since nerves control both motor and sensory functions, damage leads to multiple symptoms:**

**1. Muscle Weakness & Atrophy: Due to impaired nerve signal transmission to muscles, leading to muscle wasting.**

**2. Sensory Loss: Patients may lose their ability to feel touch, pain, or temperature in affected areas.**

**3. Pain: Nerve damage can cause chronic pain, which may feel like burning, sharp, or electric shocks.**

**4. Paresthesia: Abnormal sensations like numbness, tingling, pricking, or burning without an actual physical cause.**

**5. Autonomic Dysfunction (in severe cases): If autonomic nerves are affected, it can lead to issues such as: Loss of bowel and bladder control, Blood pressure instability and Abnormal sweating.**

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**Peripheral neuropathy refers to damage or dysfunction of one or more peripheral nerves, leading to impaired nerve function.**

**Peripheral neuropathies are classified based on the underlying mechanism of nerve damage:**

1. **Axonal Neuropathy: Involves degeneration of the axon itself and Most common type (80-90% of cases), Caused by conditions affecting nerves directly or their blood supply.**

2. **Demyelinating Neuropathy: Involves damage to the myelin sheath while the axon remains intact, Caused mainly by hereditary disorders or immune-mediated destruction of myelin.**

**Symptoms depend on whether sensory, motor, or autonomic nerves are affected:**

**Sensory Symptoms (if sensory nerves are damaged): Numbness, tingling, or burning sensations (paresthesia), Loss of sensation (touch, pain, temperature) and Increased sensitivity to pain (hyperalgesia).**

**Motor Symptoms (if motor nerves are damaged): Muscle weakness and wasting (atrophy), Loss of reflexes and Difficulty walking or performing fine motor tasks.**

**Autonomic Symptoms (if autonomic nerves are affected): Blood pressure fluctuations, Loss of bladder or bowel control and Abnormal sweating or temperature regulation issues.**

**Causes of Peripheral Neuropathy**

**1. Diabetes (Diabetic Neuropathy) – Most common cause of generalized peripheral neuropathy.**

**2. Hereditary Causes – Example: Charcot-Marie-Tooth disease.**

**3. Alcoholism – Leads to nerve damage due to toxicity and nutritional deficiencies.**

**4. Chronic Renal Failure – Toxins accumulate and damage nerves.**

**5. Neurotoxic Drugs – Chemotherapy, certain antibiotics, or heavy metals.**

**6. Autoimmune Diseases – Guillain-Barré syndrome, lupus, rheumatoid arthritis.**

**7. Nutritional Deficiencies – Vitamin B12 deficiency (essential for nerve function).**

**8. Vasculitis (Blood Vessel Inflammation) – Reduces blood supply to nerves, causing damage.**

**9. Infections – HIV, Lyme disease, leprosy, syphilis.**

**10. Tumors – Can compress nerves or infiltrate them.**

**11. Trauma or Mechanical Injury – Nerve compression or direct injury (carpal tunnel syndrome).**

**12. Amyloidosis – A disease where abnormal protein deposits damage nerves.**

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**Diabetic neuropathy is the most common complication of diabetes, caused by long-term high blood sugar (hyperglycemia) damaging peripheral nerves; the Prevalence: 7% of diabetics develop neuropathy within 1 year of diagnosis while 50% of diabetics develop neuropathy after 25 years; the Risk Factors: Duration of diabetes – the longer a person has diabetes, the higher the risk and Poor blood sugar control – higher glucose levels increase nerve damage; the Complications: Cardiovascular autonomic neuropathy shortens life expectancy and Loss**

**of sensation in the lower limbs increases the risk of foot ulcers, infections, and amputations (1–2% of diabetics require amputation).**

**Types of Diabetic Neuropathy & Clinical Manifestations:**

**Diabetic neuropathy can affect multiple nerves (polyneuropathy) or a single nerve (mononeuropathy).**

**1. Distal Symmetric Sensorimotor Polyneuropathy (Most Common Form): Affects longest nerves first, beginning in the feet and later the hands and Causes numbness, tingling, weakness, and pain in a "stocking-glove" pattern.**

**2. Autonomic Neuropathy: Affects autonomic nervous system, leading to: Gastrointestinal issues (constipation, diarrhea), Bladder dysfunction (urinary retention, incontinence) and Heart problems (irregular heartbeat, low blood pressure, fainting).**

**3. Lumbosacral Neuropathy: Affects nerves in the lower spine, leading to pain in the lower legs, weakness, and muscle wasting.**

**Symptoms of Peripheral Diabetic Neuropathy**

**Sensory Symptoms: Numbness or loss of sensation (difficulty feeling pain or temperature changes), Tingling, burning, or sharp pain and Increased sensitivity to touch (even light pressure, like a bedsheet, can cause pain).**

**Motor Symptoms: Muscle weakness, Loss of reflexes, especially at the ankle and Loss of balance and coordination (higher risk of falls).**

**Serious Complications: Foot ulcers and infections (due to numbness and poor healing) and Charcot foot – a severe deformity caused by joint and bone damage.**

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**Guillian Barre syndrome is an autoimmune neuropathy, where the body's immune system mistakenly attacks the peripheral nerves, leading to muscle weakness and paralysis, It often occurs after an infection, vaccination, or surgery.**
**Causes & Triggers: Commonly follows bacterial or viral infections, especially: Campylobacter jejuni (most common bacterial trigger), Cytomegalovirus (CMV) and Epstein-Barr virus (EBV) also Can occur after immunization or surgery.**

**CSF Findings (Cerebrospinal Fluid Analysis): Increased protein levels and Few or no white blood cells (WBCs) → This is an important diagnostic feature.**

**Clinical Features of Guillain-Barré Syndrome:**

**1. Acute Symmetric Neuromuscular Paralysis: Starts in the legs (distal) and gradually moves upward (proximal) → called ascending paralysis and Reflexes are absent (areflexia) in affected muscles.**

**2. Sensory & Autonomic Dysfunction: Some patients experience numbness, tingling, or pain and Autonomic involvement can cause: Irregular heartbeat (arrhythmia) and Blood pressure changes (hypotension or hypertension).**

**3. Fisher Syndrome (Miller-Fisher Variant of GBS): Occurs in ~5% of cases and includes: Ophthalmoplegia (eye muscle weakness), Ataxia (lack of coordination) and Areflexia (loss of reflexes).**

**4. Respiratory Failure (Severe Cases): Muscle paralysis may affect breathing, leading to respiratory failure If untreated, this can be life-threatening.**

**Course & Recovery: Symptoms peak within 2–4 weeks after onset and Most patients recover fully, but recovery can take weeks to months but Severe cases may have long-term weakness or nerve damage.**

**GBS & COVID-19: Recent studies have linked COVID-19 infection to Guillain-Barré syndrome, COVID-19 may trigger an abnormal immune response, leading to nerve damage; Patients recovering from COVID-19 should be monitored for neurological symptoms like weakness, tingling, or paralysis.**

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**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP): is a chronic, immune-mediated polyneuropathy that causes progressive or relapsing sensorimotor nerve damage for 2 months or more, It is similar to Guillain-Barré syndrome (GBS) but is chronic and progressive rather than acute; the Key Characteristics are: Mixed Sensorimotor Polyneuropathy → affects both sensory and motor nerves and Immune-mediated, but no clear history of infection (unlike GBS) and Associated with other autoimmune diseases (lupus, rheumatoid arthritis) also More common in AIDS patients due to immune dysfunction.**

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**All of the following statements are correct regarding diabetic neuropathy except:**
**A. Is the most common cause of peripheral neuropathy.**
**B. distal symmetric sensorimotor polyneuropathy is the most common form of**
**diabetic neuropathy**
**C. Can be mononeuropathic (affecting one nerve)**
**D. Autonomic nerves are never affected.**

**The correct answer is:D**

**Explanation:**

**A is correct: Diabetic neuropathy is the most common cause of peripheral neuropathy.**

**B is correct: Distal symmetric sensorimotor polyneuropathy is indeed the most common form of diabetic neuropathy.**

**C is correct: Diabetic neuropathy can also present as mononeuropathy (affecting one nerve), but polyneuropathy is more common.**

**D is incorrect: Autonomic nerves can be affected in diabetic neuropathy, leading to issues like cardiovascular problems, bladder dysfunction, and gastrointestinal issues. Therefore, autonomic nerves can indeed be affected.**

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In neurosurgery, it's not enough to understand anatomy; you must also understand how tissue dies and how it fights to survive. Every tumor, every hemorrhage, every microscopic injury carries a story in pathology - and your story as a neurosurgeon is written when you master reading it.