TRANSVERSE MYELITIS

Dhivakar Murugan

MPT (Neurology)

Assistant Professor

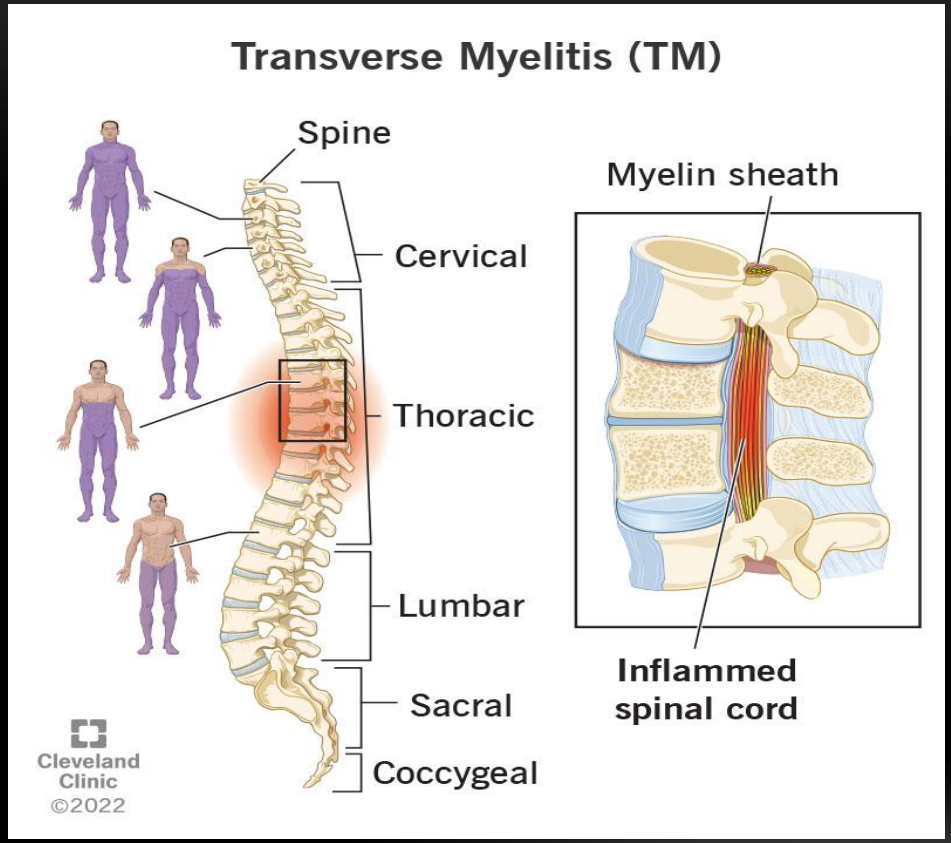
SYNOPSIS:

- Definition
- Epidemiology
- Etiology
- Types of TM
- Pathophysiology
- Clinical Presentation
- Diagnosis
- Differential Diagnosis
- Medical Management

DEFINITION

Transverse Myelitis (TM) is a clinical syndrome in which an immune-mediated process causes neural injury to the spinal cord, resulting in varying degrees of weakness, sensory alterations and autonomic dysfunction.

The word myelitis is derived from Greek words ‘myelos’ (spinal cord) and ‘itis’ (inflammation). ‘Transverse’ refers to the inflammation being across the width of the spinal cord.


Transverse Myelitis (TM)
Spine
Cervical
Thoracic
Lumbar
Sacral
Coccygeal
Myelin sheath
Inflammed spinal cord
Cleveland Clinic
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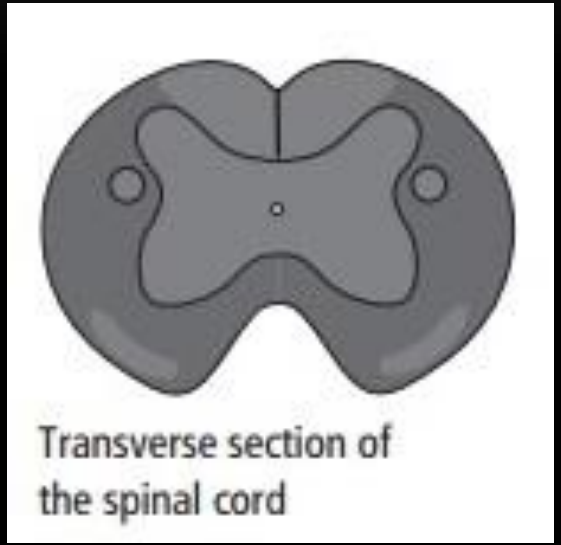

Transverse section of the spinal cord

EPIDEMIOLOGY

- Incidence between 1 – 8 new cases per million per year.
- TM affects individuals of all ages with bimodal peaks between the ages of 10 and 19 years and 30 and 39 years.
- There is no sex or familial predisposition to TM.

ETIOLOGY

The causes of transverse myelitis is categorised into following three types:

- Idiopathic
- Infectious
- Non-infectious

❖ **IDIOPATHIC:**

The clinical events are consistent with transverse myelitis that have no identifiable underlying cause.

❖ INFECTIOUS:

- **Viral Cause:**
  - Entero Virus
  - Herpes Zoster
  - Myelitis of AIDS
  - Herpes Simplex
  - Epstein bar Virus
  - Cytomegalo Virus
  - Rabies
- **Bacterial/Fungal/Parasitic:**
  - Mycoplasma Pneumonia
  - Pyogenic Myelitis
  - Tuberculous Myelitis
  - Syphilitis Myelitis
  - Epidural abscess
  - Spinal cord abscess
  - Parasitic and fungal infection producing epidural granuloma.

- **NON-INFECTIOUS:**
  - Post vaccinal myelitis
  - Acute and chronic relapsing or progressive MS
  - Sub-acute necrotizing myelitis
  - Myelopathy with lupus
  - Para neoplastic myelopathy.

TYPES OF TM

- **COMPLETE TM:**
  - Complete involvement of the spinal cord segment may leads to severe motor and sensory paralysis.
  - Usually longer (more than 3 segments involved).
- **INCOMPLETE TM:**
  - Partial or section of spinal cord is involved.
  - Usually shorter (Less than 3 segments involved).

Classification based on the spinal cord lesion characteristics:

- **ACUTE PARTIAL TM:**
  - Asymmetrical lesion involved in 1 or 2 vertebral segment.
  - Usually mild to moderate in severity.
  - Symptoms may develops over minutes to hours.

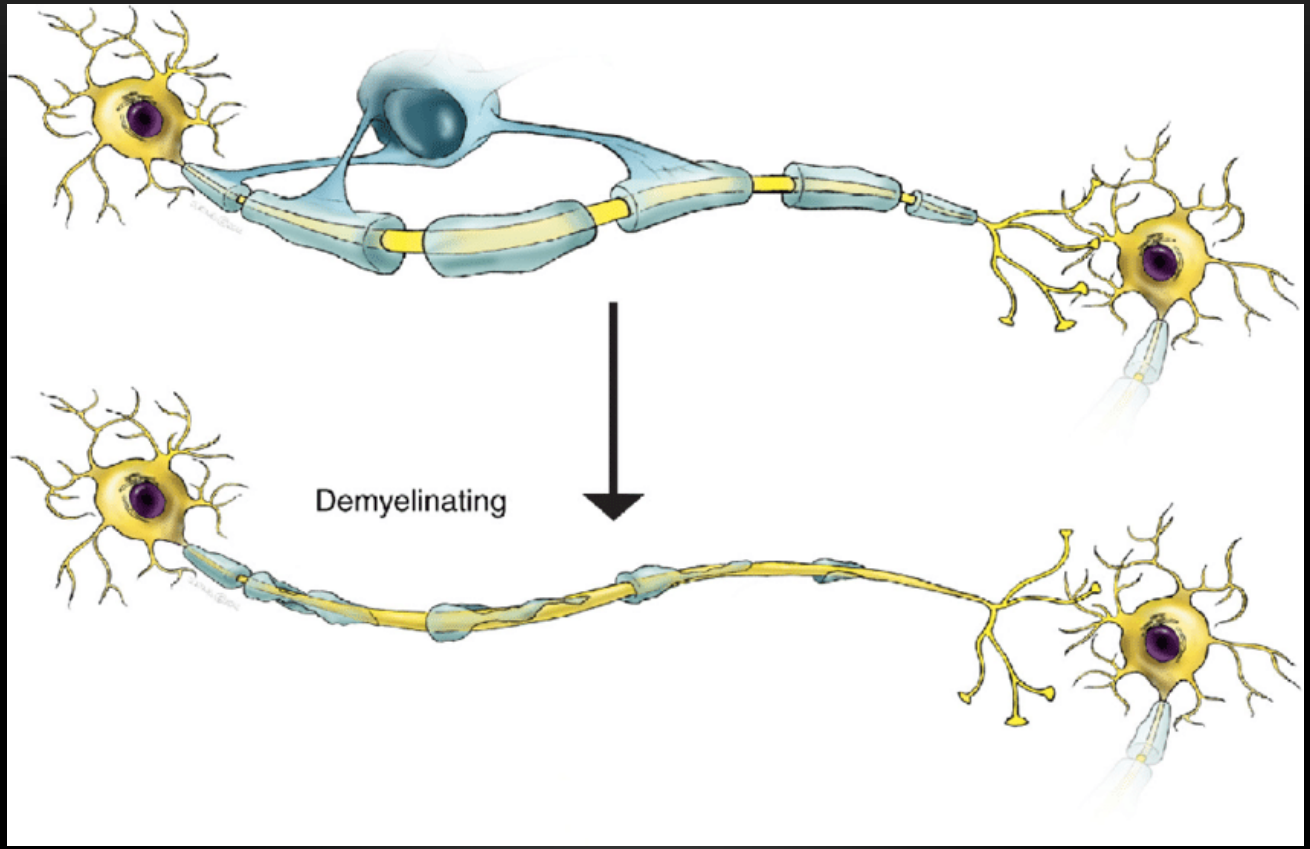
❖ LONGITUDINALLY EXTENSIVE TM:

- Symmetrical lesion of spinal cord involved on more than 3 vertebral segment.
- Usually Moderate to severe
- Symptoms may develops over days to weeks.

PATHOPHYSIOLOGY

- It is not a result of direct effect of virus on the spinal cord. But it is due to an autoimmune response of the body which provoked by infection.
- The lesions are usually due to inflammation. Involvement of spinal cord is usually central, uniform and symmetric which typically affect the cord in a patchy way.

- There is a lesion in both white and grey matter of the spinal cord which appears oedematous, hyperemic and infiltrated by inflammatory cells.
- Demyelination of neuron in the affected spinal cord may occurs due to the following three factor:
    - Cell mediated autoimmune response
    - Autoimmune vasculitis
    - Direct viral invasion of the spinal cord


Demyelinating

CLINICAL PRESENTATION

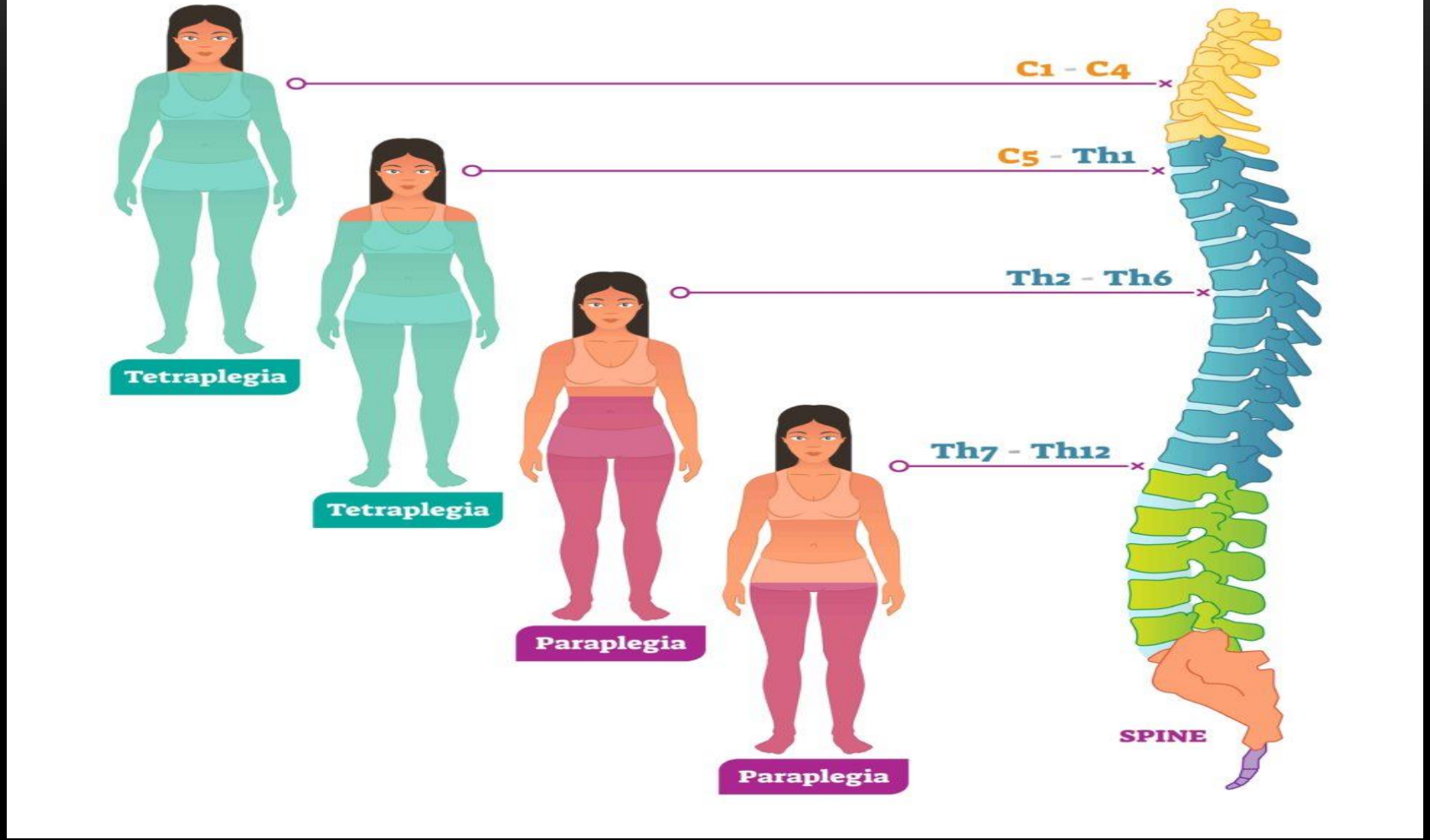
Features of transverse myelitis may develop rapidly over several hours. 37% of patients worsen maximally within 24hrs.

The clinical presentation of TM is categorised into 4 groups:

- Motor Symptoms
- Sensory Symptoms
- Autonomic
- Pain

- **MOTOR SYMPTOMS:**
  - The myelitis of the neurons of spinal cord may leads to rapid progressive weakness of muscles of leg and arm.
  - The symptoms are develops with paraparesis and may progress to paralysis.
  - Hypertonicity
  - Exaggerated reflexes
  - Positive Babinski sign.

- Paralysis is depends upon the involvement of the spinal cord.
  - Cervical level – Quadriplegia, Respiratory failure
  - Thoracic level – Paraplegia
  - Lumbar level - Paraplegia
- Spinal shock


C1 - C4
C5 - Th1
Th2 - Th6
Th7 - Th12
Tetraplegia
Tetraplegia
Paraplegia
Paraplegia
SPINE

- **SENSORY SYMPTOMS:**
  - Paraesthesia
  - Numbness
  - Altered sensation
  - Lhermitte Sign
  - Loss of pain and temperature
- **AUTONOMIC SYMPTOMS:**
  - Bladder: Urinary urgency, Incontinence, Nocturia
  - Bowel: Incontinence, Constipation
  - Sexual Dysfunction

- **PAIN:**
  - Initially - Neck and back pain
  - Later – Radiating to the extremity
  - Banding pain
  - Burning pain

DIAGNOSIS CRITERIA OF TM

❖ **Inclusion Criteria:**

- Development of sensory, motor or autonomic dysfunction attributable to the spinal cord
- Bilateral signs and/or symptoms
- Clearly-defined sensory level
- Inflammation within the spinal cord demonstrated by CSF pleocytosis *or* elevated IgG index

❖ Exclusion Criteria:

- History of previous radiation to the spine within the past 10 years
- Evidence of thrombosis of the anterior spinal artery
- Evidence of Extra-axial compressive etiology by neuroimaging.
- Abnormal flow voids on the surface of the spinal cord c/w AVM

- Serologic or clinical evidence of connective tissue disease (sarcoidosis, Behcet's disease, Sjogren's syndrome, SLE, mixed connective tissue disorder.
- History of clinically apparent optic neuritis
- CNS manifestations of syphilis, Lyme disease, HIV, HTLV-1, mycoplasma, other viral infection.
- Brain and spinal cord MRI abnormalities suggestive of MS and presence of oligoclonal bands in CSF

INVESTIGATION

Diagnosis of TM is based on the clinical and radiological findings:

- **CSF Findings:**

  - Elevated protein (100-120mg/100ml)

  - Moderate lymphocytosis (<100/mm^3)

  - Normal glucose level

  - Absence of oligoclonal bands.

- MRI Spine:

  - Enlargement of spinal cord involved in more than 3 to 4 segments.

  - Lesion occupies greater than 2/3rd of the cross sectional area of the spinal cord.

DIFFERENTIAL DIAGNOSIS

- Demyelinating disorders of spinal cord
- Other Inflammatory disease of Spinal cord
- GBS
- Multiple Sclerosis
- Compression of spinal cord due to spinal abscess.

MEDICAL MANAGEMENT

- Methylprednisolone (30mg/kg) IV for 3-7 Days
- Corticosteroids
- Intravenous Immunoglobulin
- Rituximab
- Cyclophosphamide
- **Plasma Exchange (PLEX) -** It is often initiated if a patient has moderate to severe TM.

PROGNOSIS

- Patient with idiopathic TM have at least a partial recovery, which usually begins within one to three month.
- Only 40% of patients suffering due to persistent disability.
- It is monophasic illness. Only small percentage of patients may suffer a recurrence.
- Rapid onset with complete paraplegia have been associated with poorer outcomes.