ACUTE FLACCID PARALYSIS

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PATIENT PROFILE

- 7 year / female
- Developmentally normal / No significant illness in the past
- Chief complaints : **History of acute onset of weakness of both legs followed by weakness of both arms and inability to stand or walk for 1 day.**
- No history of fever or headache or vomiting or seizures or altered sensorium or visual disturbances.
- No history of bladder or bowel involvement.
- No history of trauma/ recent vaccination/ intramuscular injection/ dog bite
- No history of contact with Tuberculosis.

CLINICAL EXAMINATION

- Alert
- Weight : 19.4kg
- Heart rate : 98/minute
- Respiratory rate : 30/minute, **Shallow respiration**
- SpO2: 99% in room air
- Blood pressure: Normal

- No neurocutaneous markers

NERVOUS SYSTEM EXAMINATION

- Higher mental function : Normal

- No ophthalmoplegia; EOM- Normal; No facial deviation
- **Gag reflex – poor**

- **Hypotonia +**
- **Power in both lower limbs -1/5**
- **Power in both upper limb - 3/5**
- **DTR – not elicitable (Areflexia)**
- B/l Plantar – withdrawal

- Spine – no deformity or tenderness

ACUTE FLACCID PARALYSIS!!

- Ascending paralysis – rapidly progressing
- Symmetric weakness
- Areflexia
- Shallow respiration
- **No bowel and bladder involvement**

- **PROVISIONAL DIAGNOSIS : GBS**
- **Plan : IVIG**

Table 607-4	Differential Diagnosis of Acute Flaccid Paralysis

Brainstem stroke
Brainstem encephalitis
Acute anterior poliomyelitis
- Caused by poliovirus
- Caused by other neurotropic viruses

Acute myelopathy
- Space-occupying lesions
- Acute transverse myelitis

Peripheral neuropathy
- Guillain-Barré syndrome
- Post–rabies vaccine neuropathy
- Diphtheritic neuropathy
- Heavy metals, biologic toxins, or drug intoxication
- Acute intermittent porphyria
- Vasculitic neuropathy
- Critical illness neuropathy
- Lymphomatous neuropathy

Disorders of neuromuscular transmission
- Myasthenia gravis
- Biologic or industrial toxins
- Tic paralysis

Disorders of muscle
- Hypokalemia
- Hypophosphatemia
- Inflammatory myopathy
- Acute rhabdomyolysis
- Trichinosis
- Familial periodic paralyses (normokalemic, hypokalemic, hyperkalemic)

From Hughes RAC, Camblath DR: Guillain-Barré syndrome, Lancet *366:1653–1666, 2005.*

SHIFTED TO PICU

- In view of respiratory involvement - possibility of requiring NIV

BEFORE STARTING IVIG

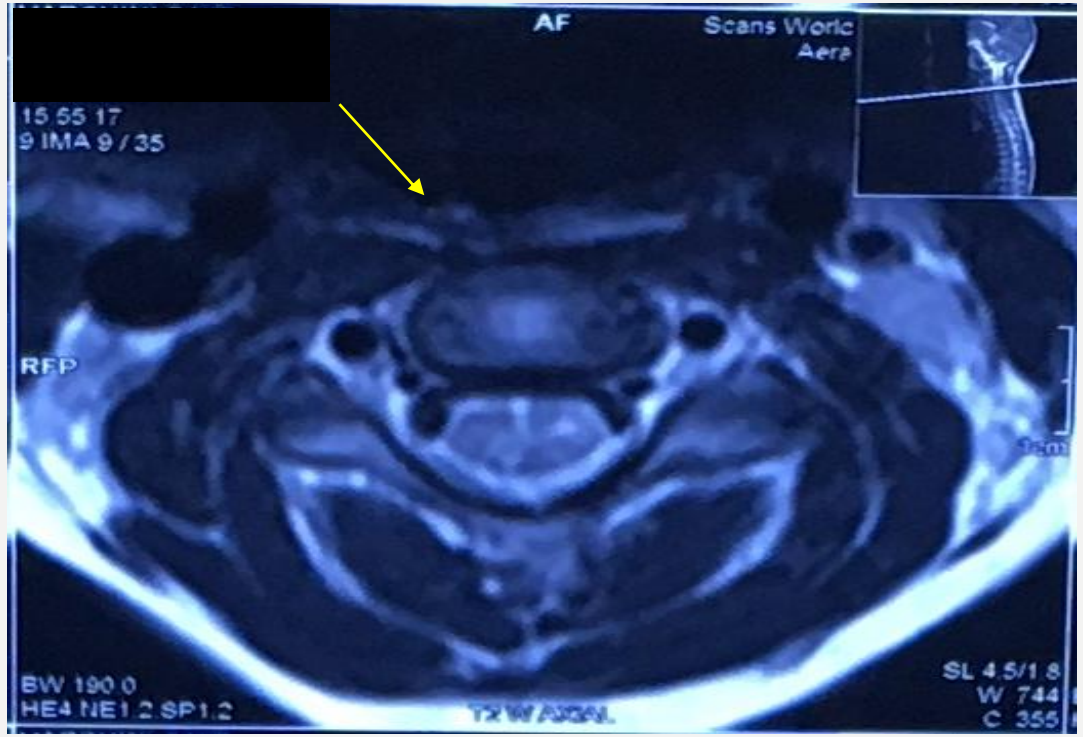
Sensory examination : Hyperalgesia over 2nd intercostal space

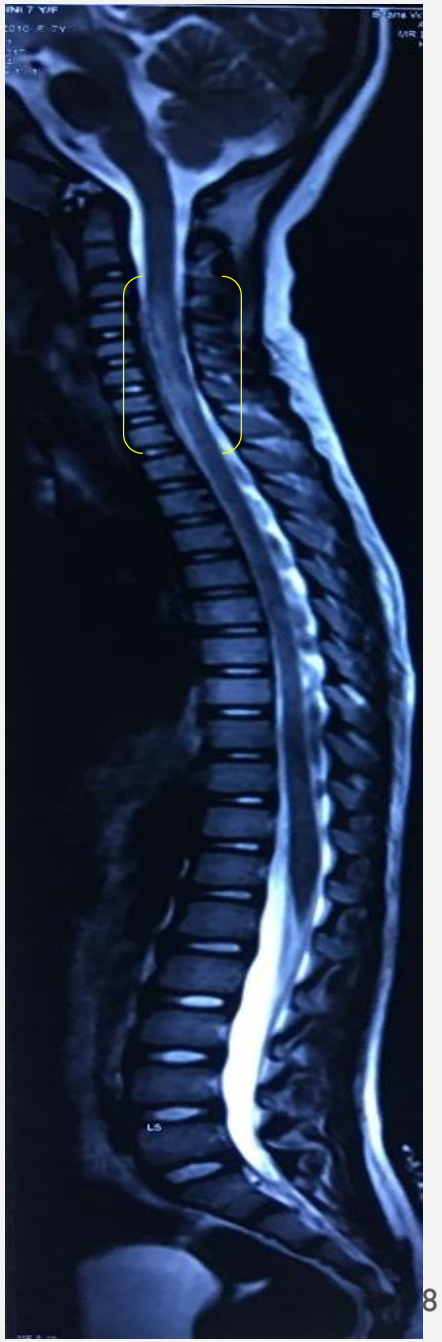
Plan : MRI brain and spine

NEUROIMAGING

- MRI spine showed mild cord swelling with altered intensity from **C3 to T1** level **involving more than 2/3rd of the cord at C4 level** (features suggestive of **acute demyelination of spinal cord** at C3 to T1 level).

- MRI Brain : Normal





INVESTIGATIONS

- **CSF analysis: protein 12 mg/dl, sugar 73 mg/dl, 4 lymphocytes**
- **CSF viral panel : Negative**
- Hb: 7.1 g/dL; TC : 10,400; N55 L40; PCV : 23.4%, Platelet : 2.92 Lac
- Renal Function: Normal

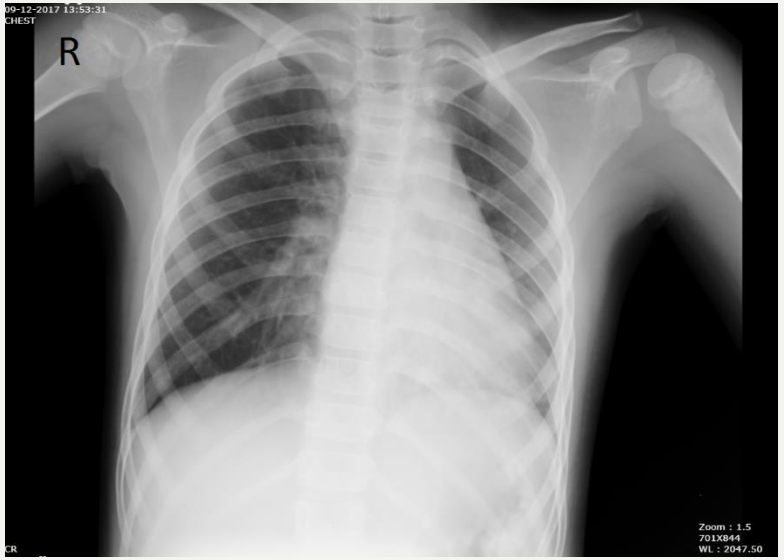
LONGITUDINALLY EXTENSIVE TRANSVERSE MYEITIS

- No evidence of Optic Neuritis
- Started on Pulse Methylprednisolone

COURSE IN THE PICU

- Urinary retention – catheterised
- Collapse left lung – NIV

- Completed 5 days of MPS



FURTHER COURSE (4 WEEKS OF PICU)

- Worsening collapse
- Paradoxical breathing – requiring NIV

- Second line therapy
- 9 cycles of **Plasmapheresis**

- Hospital Acquired Infection (blood stream & urinary)
- Thrombosis secondary to cannula

GRADUAL IMPROVEMENT

- B/L Upper limbs power improved to 4 /5
- Left lower limb power improved to 3 /5
- Right lower limb power improved to 2/5
- Bladder control+

WORK UP:

- **Serum NMO antibody – negative**
- **Thyroid profile – normal**
- **Vasculitis – workup planned**

Table 600-1	Differential Diagnosis of Demyelinating Disorders

Acute disseminated encephalomyelitis (ADEM)
Multiple sclerosis (including tumefactive MS)
Acute hemorrhagic leukoencephalopathy
Clinically isolated syndrome (CIS)
Neuromyelitis optica spectrum disorder
N-methyl-D-aspartate receptor (NMDAR) antibody and other autoimmune encephalitis
Vasculitis/angiopathies
Hashimoto encephalitis (anti–thyroid peroxidase [TPO] antibody)
Familial hemophagocytic lymphohistiocytosis
Langerhans cell histiocytosis
Lymphoma
Gliomatosis cerebri
Glioma
Sarcoidosis
Mitochondrial disorders (Leigh syndrome)
Vitamin E deficiency
Vitamin B_{12} deficiency
Celiac disease
Herpes simplex virus (HSV), enterovirus, arbovirus, Powassan and other viral encephalitides
Rabies
Subacute sclerosing pan-encephalitis (SSPE) (chronic measles)
Charcot-Marie-Tooth syndrome
Leukoencephalopathies (Aicardi-Goutières syndrome)
Vanishing white matter disease
Schilder disease (possibly an adrenoleukodystrophy)
X-linked adrenoleukodystrophy
Griscelli syndrome type 2

FOLLOW UP (2 WEEKS AFTER DISCHARGE)

- B/L Upper limb power : 4 /5
- B/L Lower limb power : 3 /5
- Weak hand grip +
- Clawing of B/L hand
- Right foot drop
- Brisk DTR
- Bowel and bladder : N

Table 3 Characteristics to aid differential diagnosis of acute flaccid paralysis

Feature	Transverse myelitis	Poliomyelitis	Guillain-Barre syndrome	Traumatic neuritis (following injection)
Development of paralysis	From hours to four days	24 to 48 h from onset to full paralysis	From hours to 4 wk	From hours to four days
Fever at onset of weakness	May be present	High, always present at onset of flaccid paralysis	Uncommon	Present, if underlying infection being treated with IM injections
Paralysis	Symmetric	Asymmetric,	Symmetric, mostly ascending	Affects only one limb
Progression of paralysis		Descending•	Ascending	
Muscle tone	Reduced during acute phase	Reduced	Reduced	Reduced
Deep-tendon reflexes	Absent in lower limbs(early); hyperreflexia(late)	Decreased or absent	Absent	Decreased or absent
Sensation	Anesthesia of lower limbs with sensory level	Severe myalgia, backache, no sensory changes	Cramps, tingling, hypoanesthesia of palms and soles	Pain in gluteus
Cranial nerve involvement	Absent	Only when bulbar involvement is present	Often present, affecting nerves VII, IX, X, XI, XII	Absent
Respiratory insufficiency	Sometimes	Only when bulbar involvement is present	Occurs in severe cases	Absent
Autonomic signs and symptoms	Present	Rare	Frequent in severe cases (blood pressure alterations, sweating, blushing, and body temperature fluctuations)	Hypothermia in affected limb
Cerebrospinal fluid	Normal or Pleocytosis	Mild elevation of lymphocytes 10 to 200/mL	Albumin-cytologic dissociation (usually <10 cells/ml, never >50cells/ml)	Normal
Bladder dysfunction	Present- early and persistent	Rare	Occasionally (Transient, at the peak of weakness, 1–3 d (30 %))	Never
Nerve conduction velocity: third wk	Normal	Abnormal: anterior horn cell disease (normal during first 2 wk)	Abnormal: slowed conduction, decreased motor amplitudes	Abnormal: s/o motor-sensory axonal damage
Diagnostic test	MRI–spine	Stool viral detection	Nerve conduction studies	Nerve conduction studies, Electromyography

Adapted and modified from Global Program for Vaccines and Immunization: Field Guide for Supplementary Activities Aimed at Achieving Polio Eradication. Geneva, World Health Organization, 1996

LETM

- LETM is spinal cord lesion that extends over **three or more vertebrae,** causing **T2 hyperintensity** on MRI spine.
- LETM is a characteristic feature of **NMO.**
- Treatment hinges on distinguishing inflammatory and non inflammatory aetiologies and identifying patients who are at a high risk of a recurrent course.

LETM – DIFFERENTIAL DIAGNOSIS

NMO
NMO spectrum

Inflammatory
MS
ADEM
SLE, Sjogren, Behcet

Infectious
Parainfectious

Neoplastic
Trauma

Nutritional

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Mimics and chameleons in Guillain–Barré and Miller Fisher syndromes

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