

IndSPN Case of the Month

January 2023

Clinical Presentation

A eight-year-old girl born out of non-consanguineous marriage brought by parents with chief complaints of-

- Tuft of hair over mid-back region since birth
- Constipation for past 4 years
- Bilateral lower limb weakness for past 3 years

Clinical Examination

- Child was alert and playful
- Higher mental functions- Intact
- Nutrition– Adequate
- Tone– Normal in all 4 limbs
- Power– 5/5 in all four limbs except bilateral ankle 4/5
- Deep tendon reflexes– B/L ankle jerk 1+, rest 2+
- Plantar- B/L mute

Clinical Imaging



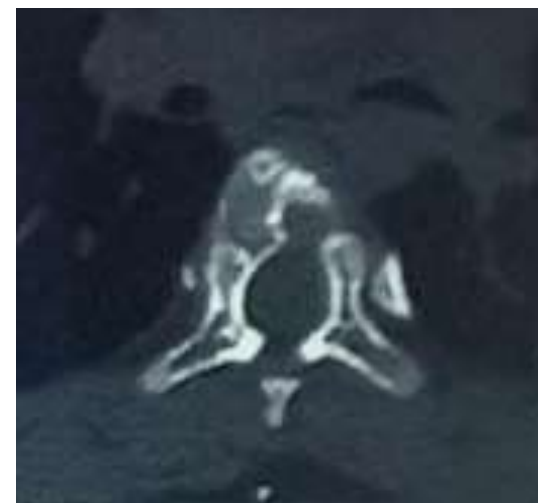
Tuft of hair at mid thoracic region

Provisional Diagnosis with Clinical Localization



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Radiology



➤ *MRI Spine (Plain + Contrast)*

- There is evidence of short segment syringomyelia at D7 vertebrae. The spinal cord is terminating at L1-L2 level. Filum terminale is thickened

➤ *MRI Brain*

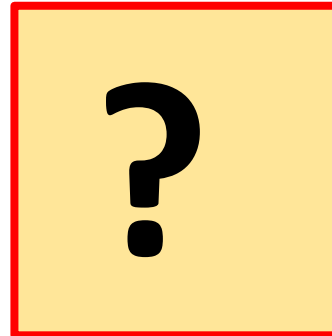
- There is no evidence of tonsillar herniation/hydrocephalus

➤ *CT Spine*

- Evidence of butterfly vertebrae at D5 level

Radiological Impression

➤ **Differential Diagnosis**



Surgery

- **Position-** Prone position
- **Incision-** Midline skin incision extending from L4 to S1
- **Procedure-** L5 partial laminectomy and detethering of cord under triggered EMG monitoring

Intraoperative Images

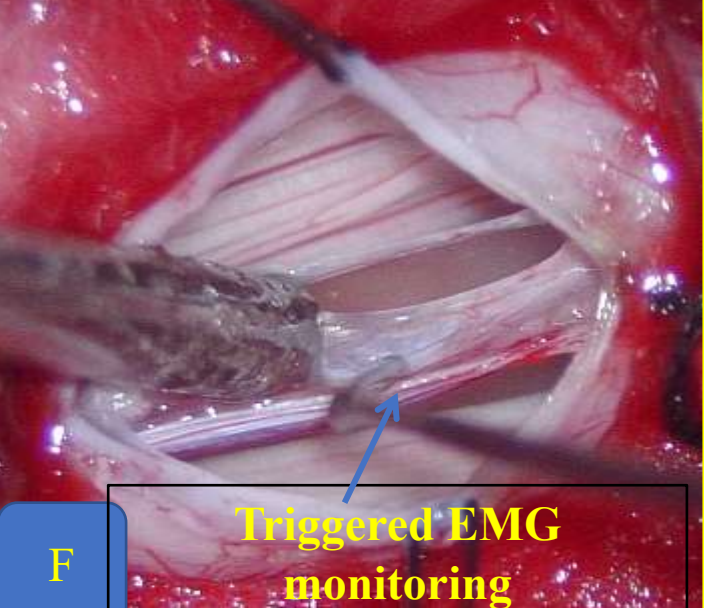
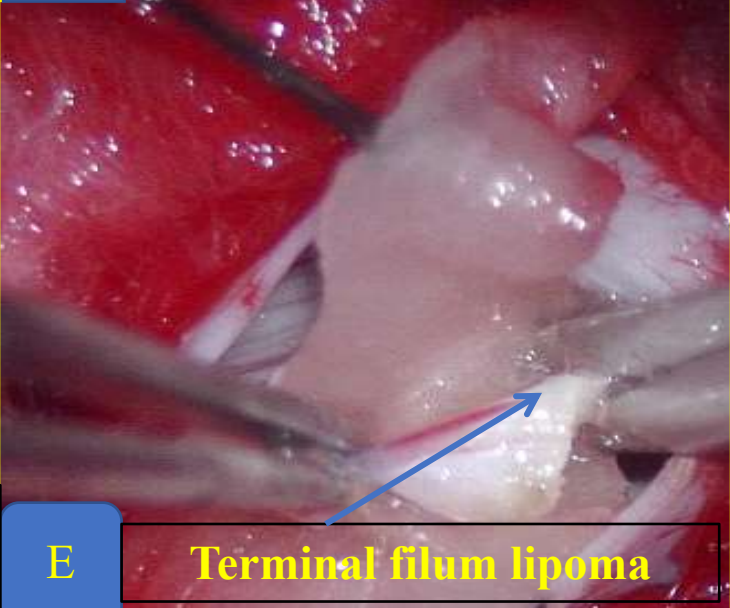
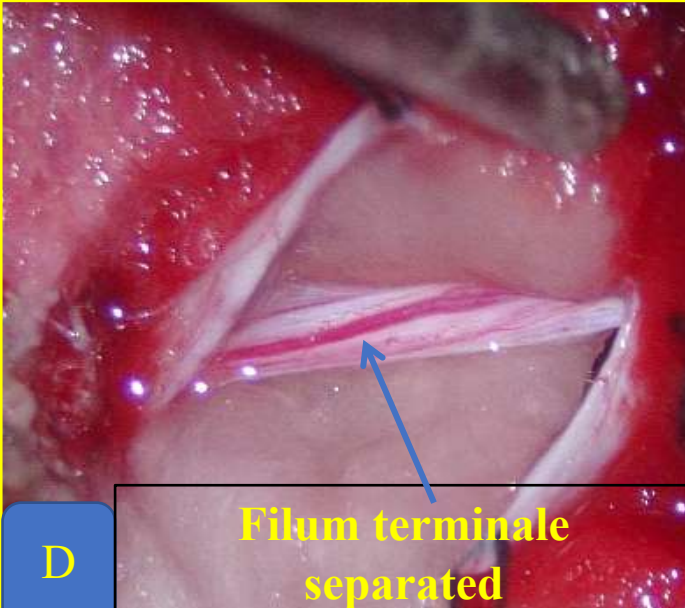
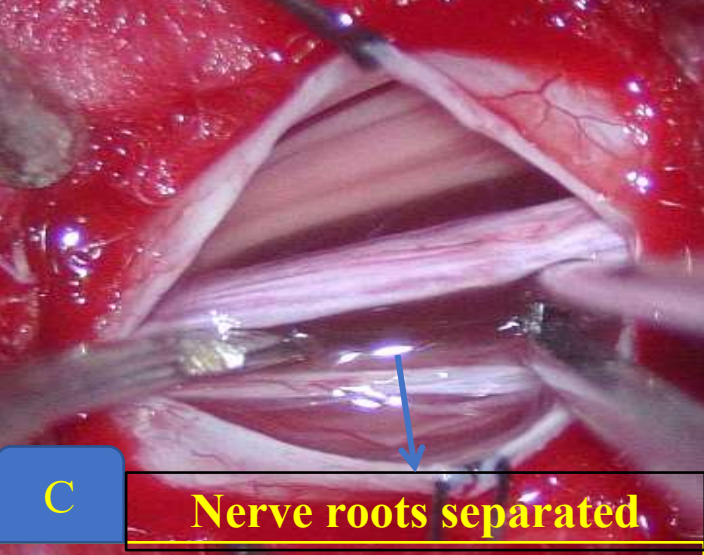
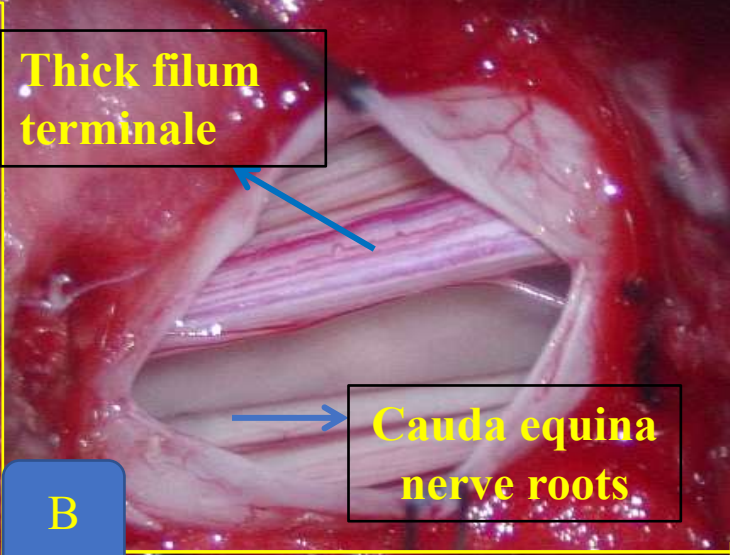
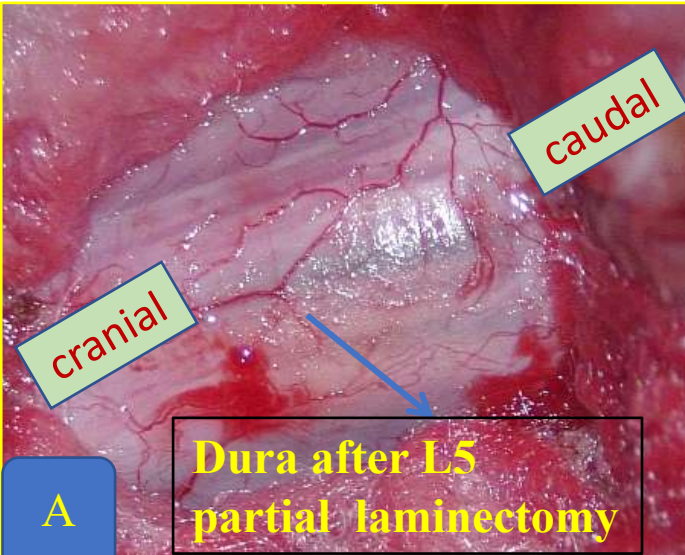


Patient Positioning

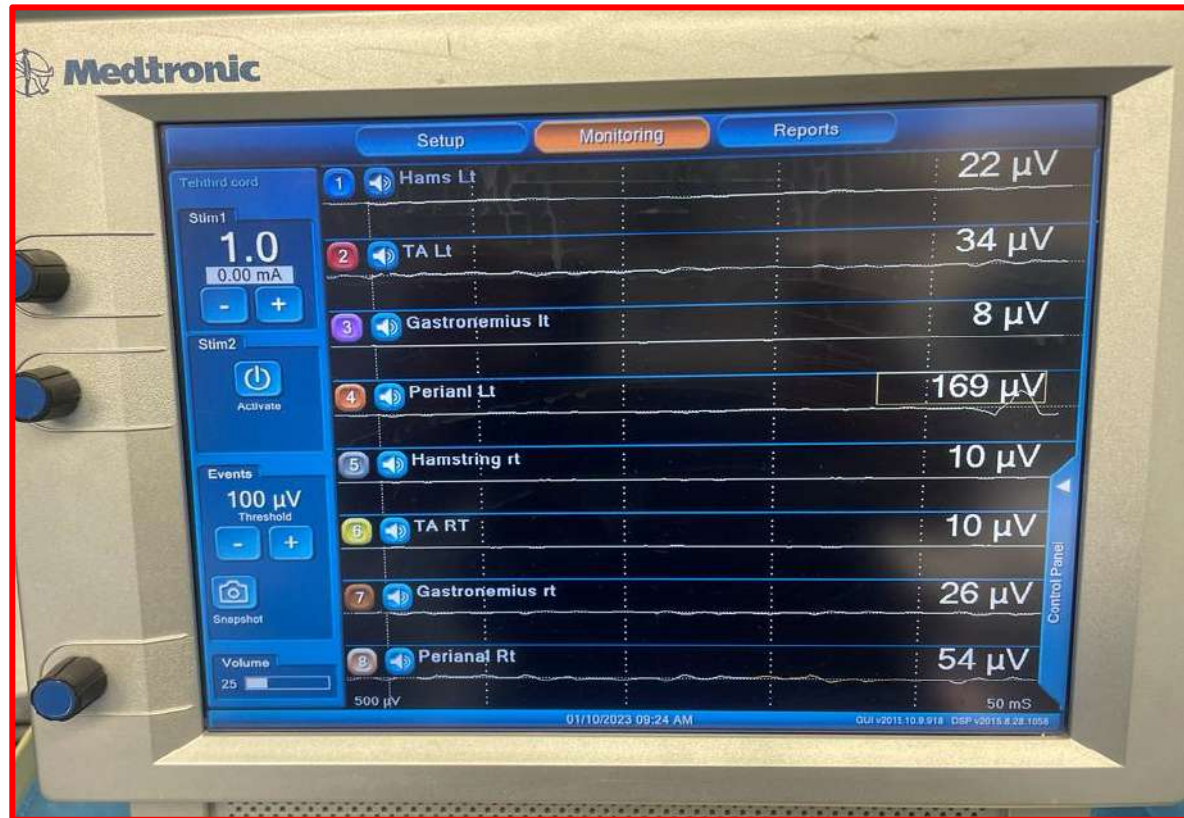


Baseline EMG Recording

Intraoperative Images



Intraoperative Neuromonitoring Recording



Triggered EMG tracings showing neuronal integrity

Post-operative status



Healthy wound

- Improvement in Power
- No bowel and bladder complaints

Case Summary

- This 8-year-old girl was brought with chief complaints of tuft of hair over mid-back region since birth with constipation and bilateral lower limb weakness for past 3 years. Radiological evaluation was suggestive of short segment syringomyelia at D7 with cord terminating at L1-L2 level with a thickened filum terminale
- L5 partial laminectomy and detethering of the cord was done. Filum terminale was cut under EMG monitoring
- Post-operatively there was improvement in power

Relevant Literature

- Tethered cord syndrome refers to a group of neurological disorders that result from chronic traction to spinal cord as a consequence of it being “tethered” or held by various structures
- It is a component of “occult spinal dysraphism”
- A taut (short fibrous) or a thick (fat infiltrated) filum is inelastic and causes excessive traction on the conus during spinal movements
- Cord suffers repeated mechanical shocks if its movement is restricted by fixity to a dorsal (e.g. lumbosacral lipoma) or ventral lesion (e.g., bony spur)

- Tethered cord causes ischemia and metabolic changes that impair neural transmission resulting in neuronal dysfunction. Eventually, tethering of the cord can produce irreversible neuronal damage that is seen histologically
- MRI which demonstrated conus ending way below the normal L1–L2 junction or had fat signals in the filum causing a thickened filum which was defined as more than 2 mm in diameter
- Treatment includes surgical detethering including removal of the offending agents such as fat, bony spur, sinus tracts etc.

Suggested Readings

- Tandon V, Garg K, Kumar R, Mahapatra AK, Sharma BS. Management of adult tethered cord syndrome: Our experience and review of literature. *Neurology India* 2014;62:137-43.
- Vepakomma D, Kumar N, Alladi A. Tethered Cord Syndrome-Role of Early Surgery. *J Indian Assoc Pediatr Surg.* 2019;24:124-8.
- Pang D. Perspectives on Spinal Dysraphism: Past, Present, and Future. *J Korean Neurosurg Soc.* 2020;63:366-72.