A confounding pediatric spinal cord injury: anterior, central, or both?

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Context

Pediatric spinal cord injury (SCI) most commonly affects the cervical region. Central cord syndrome (CCS) most often occurs in the lower cervical spinal cord due to hyperextension injury, while anterior cord syndrome is primarily due to vascular infarction after hyperextension injury. We present an unusual case of a pediatric patient that physically presents with CCS but clinically has evidence of anterior spinal artery syndrome (ASAS).

Findings

Acute care presentation:

- 2-year-old male with no prior medical history and no developmental delay presented to acute care after an unwitnessed fall while jumping from approximately three feet high
- Initial physical exam
  - Bilateral upper extremity flaccid paraplegia with areflexia and dysesthesia to light touch
  - Spontaneous antigravity movement of bilateral lower extremities
  - Intact cranial nerve function, including swallowing
  - Bowel and bladder function difficult to gauge due to lack of toilet training

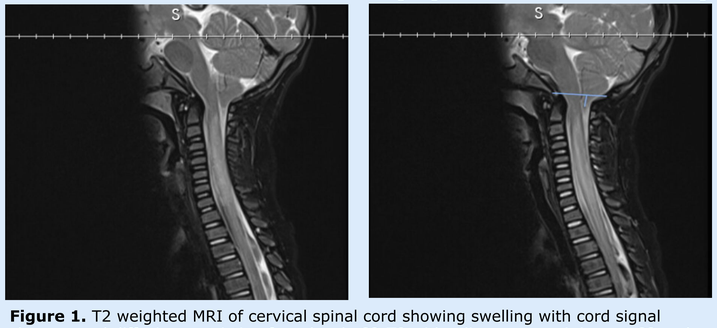


Figure 1. T2 weighted MRI of cervical spinal cord showing swelling with cord signal changes and diffusion restriction from levels C2-T3 with a ligamentous injury at C3 and Chiari I malformation.

- Radiographic interpretation
  - Imaging suggestive of anterior spinal artery (ASA) infarction in the setting of the anatomic predisposition of Chiari I
- Further workup to rule out prothrombotic diseases
  - Basic coagulation labs within normal limits
  - CTA of head and neck unremarkable, ASA not adequately seen
  - Family history unremarkable
- Treatment
  - Dexamethasone for six days
  - Miami J cervical collar for six weeks
  - No acute surgical intervention

Inpatient rehabilitation (IPR) course:

- ASIA impairment scale was deferred due to age, inability to participate, and not being potty trained

- Occupational therapy (OT)
  - Interventions: Neuromuscular electrical stimulation (NMES) trialed to attempt flaccid musculature recruitment without success despite changing settings multiple times during each session
  - Upper extremity muscle recovery:
    - Trapezius (R > L) → Anterior deltoids → Wrist extensors and supination
    - Wrist supination to compensate for weak finger flexors to assist with wrist extension for a tenodesis grasp, allowing a strong grasp (R > L)
    - Shoulder hike and trunk rotation to swing arms to compensate for no elbow flexion/extension

- Physical therapy (PT)
  - Interventions targeted to develop:
    - Upper extremity protective responses
    - Improve gait and truncal coordination, given observed lower extremity weakness for his age despite antigravity capabilities, further contributing to poor balance and increased risk of fall

- Transient dysesthesias treated with Gabapentin

Table 1. Patient's muscle recovery and therapy progress.

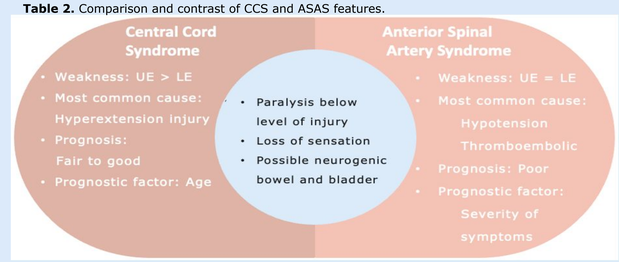
	Discharged from IPR	Six-month follow-up
Elbow flexion	- None bilaterally	- RUE: 60-70° against gravity - LUE: 50-60° against gravity
Shoulder flexion	- RUE: 10° with compensatory strategies	- RUE: 0-45° (can obtain 70° with lordosis and arm swing)
Grasping	- RUE: Using tenodesis and supination - LUE: Unable to maintain with tenodesis	- RUE: Pincer using lightweight objects
Upper extremity functionality	- Unable to carry objects	- Pick up objects with BUE to carry them - Requires max A for hand to mouth with his RUE
Supine to sit	- Mod A	- Supervision with setup of BUE
Gait	- 150ft with CGA and bilateral AFOs	- Supervision
Stairs	- Max A	- Supervision ascending and descending (scoots for safety)

***RUE**: Right upper extremity, **LUE**: Left upper extremity, **BUE**: Bilateral upper extremities
Max A: Maximal assistance, **Mod A**: Moderate assistance, **CGA**: Contact guard assist
AFOs: Ankle foot orthoses

Please use the QR code to more detailed case report and access references

Clinical relevance

Table 2. Comparison and contrast of CCS and ASAS features.



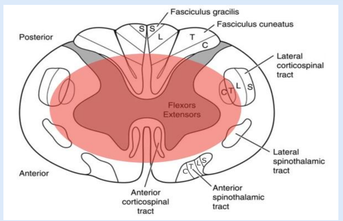


Figure 2. Simultaneous anterior and posterior spinal cord compression affects the medial aspect of the lateral corticospinal tracts in CCS.

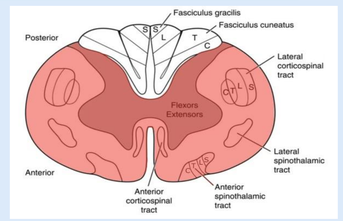


Figure 3. Loss of blood flow to the anterior spinal artery leads to involvement of the anterior 2/3 of the spinal cord, as observed in ASAS.

- Patient had physical manifestations of CCS, such as upper extremity flaccid paralysis and dysesthesias, with radiographic evidence of ASAS
- Displaying pattern typical of a lower motor neuron (LMN) SCI, but no evidence of areflexic bowel or bladder
- Recovery: CCS – follows an ascending, proximal to distal pattern
  ASAS – determined by severity of symptoms, and even with milder presentations, recovery to prior baseline is minimal
  - Patient had distal muscle recovery with minimal proximal recovery prior to discharge from IPR and improved UE functionality in six months
- Nontraumatic thromboembolism is less common in children and would most likely be associated with underlying hematologic diseases
  - Further hypercoagulable workup deferred due to likelihood of arterial ischemia secondary to Chiari I malformation
  - Two case reports found in the literature suggest Chiari I as a possible risk factor for ASA ischemia in adults but not in children
  - Further investigation is necessary in pediatric population
- In general, it is difficult to prognosticate pediatric SCI recovery. Yet it is particularly challenging to predict what the long-term functional recovery of this patient will be based on the following:
  - Clinical features of CCS with imaging signs of ASA infarct and Chiari I
  - Unusual muscle recovery pattern that was distal-to-proximal and patchy
  - Paucity of data on ASAS and CCS in children