

Spinal epidural lipomatosis and focal posterior longitudinal ligament hypertrophy causing severe cauda equina crowding

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DESCRIPTION

A male in his early fifties was referred to the local emergency department with severe lower back pain, bilateral radiculopathy and lower limb weakness. He initially presented to his general practitioner (GP) with a five-day history of bilateral radicular pain and numbness in the feet, on a background of a three-week history of worsening lower back pain. His GP referred him for MRI of the lumbar spine, which was performed two weeks later. MRI lumbar spine demonstrated a large disc extrusion producing severe cauda equina crowding with effacement of the majority of the thecal sac cerebrospinal fluid (CSF) at L3/L4 with a prominent multilayer lumbar dorsal epidural fat layer, focal posterior longitudinal ligament hypertrophy and disc protrusion at L5/S1 with moderate-to-severe bilateral foraminal stenosis (figures 1–4).

The patient was advised by the reporting radiologist to attend the on-site emergency department immediately. He was assessed by the emergency department and referred to the on-call orthopaedic surgeons. On assessment, the patient was haemodynamically stable and had no saddle anaesthesia, perianal dysaesthesia, loss of anal tone, urinary retention or faecal incontinence. His medical history was significant for hypertension and obesity. An American Spinal Injury Association (ASIA) score was performed and demonstrated multilevel neurological deficits, leading to a score of D (figure 5). He had 5/5 power in hip flexors (L2) and knee extensors (L3) bilaterally, but only 3/5 power in ankle dorsiflexion (L4) bilaterally and 4/5 power in hallux extensors (L5) and also ankle plantarflexors (S1) bilaterally. The patient also exhibited 2/2



Figure 2 Magnetic resonance sagittal image of the lumbar spine demonstrating posterior disc bulge, posterior longitudinal ligament hypertrophy and dorsal epidural fat at L3/L4 on T2.



Figure 3 Magnetic resonance sagittal image of the lumbar spine demonstrating posterior disc bulge, posterior longitudinal ligament hypertrophy and dorsal epidural fat at L3/L4 on T2.



Figure 1 Magnetic resonance sagittal image of the lumbar spine demonstrating posterior disc bulge, posterior longitudinal ligament hypertrophy and dorsal epidural fat at L3/L4 on T1.



Figure 4 Magnetic resonance axial image of the lumbar spine at L3/L4 demonstrating severe cauda root crowding on T2.



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Muscle Function Grading

- 0 = total paralysis
 1 = palpable or visible contraction
 2 = active movement, full range of motion (ROM) with gravity eliminated
 3 = active movement, full ROM against gravity
 4 = active movement, full ROM against gravity and moderate resistance in a muscle specific position
 5 = (normal) active movement, full ROM against gravity and full resistance in a functional muscle position expected from an otherwise unimpaired person
 5* = (normal) active movement, full ROM against gravity and sufficient resistance to be considered normal if identified inhibiting factors (i.e. pain, disease) were not present
 NT = not testable (i.e. due to immobilization, severe pain such that the patient cannot be graded, amputation of limb, or contracture of > 50% of the normal ROM)

Sensory Grading

- 0 = Absent
 1 = Altered, either decreased/impairment sensation or hypersensitivity
 2 = Normal
 NT = Not testable

When to Test Non-Key Muscles:

In a patient with an apparent AIS B classification, non-key muscle functions more than 3 levels below the motor level on each side should be tested to most accurately classify the injury (differentiate between AIS B and C).

Movement	Root level
Shoulder: Flexion, extension, abduction, adduction, internal and external rotation	C5
Elbow: Supination	
Elbow: Pronation	C6
Wrist: Flexion	
Finger: Flexion at proximal joint, extension.	C7
Thumb: Flexion, extension and abduction in plane of thumb	
Finger: Flexion at MCP joint	C8
Thumb: Opposition, adduction and abduction perpendicular to palm	
Finger: Abduction of the index finger	T1
Hip: Adduction	L2
Hip: External rotation	L3
Hip: Extension, abduction, internal rotation	L4
Knee: Flexion	
Ankle: Inversion and eversion	
Toe: MP and IP extension	
Hallux and Toe: DIP and PIP flexion and abduction	L5
Hallux: Adduction	S1

ASIA Impairment Scale (AIS)

A = Complete. No sensory or motor function is preserved in the sacral segments S4-5.

B = Sensory Incomplete. Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4-5 (light touch or pin prick at S4-5 or deep anal pressure) AND no motor function is preserved more than three levels below the motor level on either side of the body.

C = Motor Incomplete. Motor function is preserved at the most caudal sacral segments for voluntary anal contraction (VAC) OR the patient meets the criteria for sensory incomplete status (sensory function preserved at the most caudal sacral segments (S4-S5) by LT, PP or DAP), and has some sparing of motor function more than three levels below the ipsilateral motor level on either side of the body.
 (This includes key or non-key muscle functions to determine motor incomplete status.) For AIS C – less than half of key muscle functions below the single NLI have a muscle grade ≥ 3 .

D = Motor Incomplete. Motor incomplete status as defined above, with at least half (half or more) of key muscle functions below the single NLI having a muscle grade ≥ 3 .

E = Normal. If sensation and motor function as tested with the ISNCSCI are graded as normal in all segments, and the patient had prior deficits, then the AIS grade is E. Someone without an initial SCI does not receive an AIS grade.

Using ND: To document the sensory, motor and NLI levels, the ASIA Impairment Scale grade, and/or the zone of partial preservation (ZPP) when they are unable to be determined based on the examination results.



Steps in Classification

The following order is recommended for determining the classification of individuals with SCI.

1. Determine sensory levels for right and left sides.

The sensory level is the most caudal, intact dermatome for both pin prick and light touch sensation.

2. Determine motor levels for right and left sides.

Defined by the lowest key muscle function that has a grade of at least 3 (on supine testing), providing the key muscle functions represented by segments above that level are judged to be intact (graded as ≥ 3).
 Note: In regions where there is no myotome to test, the motor level is presumed to be the same as the sensory level, if testable motor function above that level is also normal.

3. Determine the neurological level of injury (NLI)

This refers to the most caudal segment of the cord with intact sensation and antigravity (3 or more) muscle function strength, provided that there is normal (intact) sensory and motor function rostrally respectively.
 The NLI is the most cephalad of the sensory and motor levels determined in steps 1 and 2.

4. Determine whether the injury is Complete or Incomplete.

(i.e. absence or presence of sacral sparing)
 If voluntary anal contraction = **No** AND all S4-5 sensory scores = **0** AND deep anal pressure = **No**, then injury is **Complete**.
 Otherwise, injury is **Incomplete**.

5. Determine ASIA Impairment Scale (AIS) Grade:

Is injury Complete? If YES, AIS=A and can record ZPP (lowest dermatome or myotome on each side with some preservation)

Is injury Motor Complete? If YES, AIS=B

NO
 NO
 (No-voluntary anal contraction OR motor function more than three levels below the motor level on a given side, if the patient has sensory incomplete classification)

Are at least half (half or more) of the key muscles below the neurological level of injury graded 3 or better?

NO
 YES
 AIS=C
 AIS=D

If sensation and motor function is normal in all segments, AIS=E
 Note: AIS E is used in follow-up testing when an individual with a documented SCI has recovered normal function. If at initial testing no deficits are found, the individual is neurologically intact; the ASIA Impairment Scale does not apply.

Figure 5 American Spinal Injury Association impairment scale.

sensation in L1-L4 dermatomes, but showed reduced sensation of 1/2 in L5 and S1. Lower limb reflexes and tone were normal. The patient was referred to the on-call neurosurgical team and underwent lumbar decompression surgery. His postoperative course was uneventful.

Hypertrophy of the posterior longitudinal ligament (HPLL) is a rare pathological condition that is characterised by thickening of the PLL, while spinal epidural lipomatosis (SEL) is a rare condition characterised by an overgrowth of adipose tissue in the extradural space, usually deposited in the thoracic or lumbar spine.¹ This can lead to stenosis of the central canal and compression of adjacent neural structures. First documented in 1975, the most common cause of SEL is exogenous steroid therapy, but

other causes have been shown to be idiopathic, iatrogenic or due to obesity. The pathogenesis resulting in neurological compromise still remains unclear.² SEL can present with radiculopathy, myelopathy, bladder or bowel dysfunction, claudication, motor weakness, sensory deficits or acute cauda equina syndrome. Conservative measures in the treatment of SEL are aimed to treat the underlying cause, including cessation of steroid use and weight loss. However, in patients presenting with severe symptoms, decompression surgery has proven to be effective.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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Learning points

- Spinal epidural lipomatosis (SEL) is a rare condition characterised by an overgrowth of unencapsulated adipose tissue in the extradural space.
- SEL can present with radiculopathy, myelopathy, bladder or bowel dysfunction, claudication, motor weakness, sensory deficits and rarely cauda equina syndrome.
- Conservative measures in the treatment of SEL are aimed to treat the underlying cause; however, in severe cases, surgery is indicated.

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