An Unnerving Situation

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Alice’s Adventures in Wonderland
by Lewis Carroll

Illustrations by Sir John Tenniel

Story and illustrations are in the public domain.
61-year old woman with history of:

- CKDIII
- Inflammatory breast cancer (2007) in remission
- Herpes genitalis
- Chronic lumbar back pain
- Tobacco use disorder
One last thing, doc. I was too embarrassed to say this before...
...I’m going to the bathroom when I don’t mean to.
...I’m going to the bathroom when I don’t mean to.  
Bowel and bladder incontinence
...I’m going to the bathroom when I don’t mean to.
Bowel and bladder incontinence
Perineal numbness
...I’m going to the bathroom when I don’t mean to.

Bowel and bladder incontinence
Perineal numbness
Back pain
Cauda Equina Syndrome

Tarulli AW. Disorders of the Cauda Equina. Continuum 2015; 21(1): 146-158
Defining Cauda Equina Syndrome

For a diagnosis of CES, one or more of the following must be present: (1) bladder and/or bowel dysfunction, (2) reduced sensation in the saddle area, or (3) sexual dysfunction, with possible neurologic deficit in the lower limb (motor/sensory loss, reflex change).
Differential Diagnosis

Compressive

- Disk herniation
- Fracture
- Neoplasms
- Iatrogenic injury
- Chronic lumbosacral spinal stenosis
- Infection

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Differential Diagnosis

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- Disk herniation
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Non-compressive
- Ischemia
- Infiltration
- Inflammation
- Infection

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Focused Neurologic Exam

- Motor Strength: 5/5 upper and lower extremities
- Reflexes: 2+ upper and lower extremities
- Pathologic Reflexes: absent Babinski sign and clonus
- Sensation: perianal sensation diminished on left
- Anal Sphincter: flaccid rectal tone
- Gait: normal
Problem-based History

- Chronic lumbar **back pain** with left-sided radiculopathy that was unchanged from baseline
- New-onset **acute** perineal numbness, urinary retention and constipation, then days later **urinary and fecal incontinence**, now only fecal incontinence
- 6-7 week duration of symptoms, **improving**
- No identifiable inciting event, trauma, fever, night sweats, weight loss, or IV drug use
Laboratory Data

- Na: 139 mmol/L
- K: 4.4 mmol/L
- Cl: 100 mmol/L
- CO2: 27 mmol/L
- BUN: 21 mg/dL
- Cr: 1.4 mg/dL
- Ca: 9.5 mg/dL
- AST: 28 U/L
- ALT: 18 U/L
- Alk Phos: 40 U/L
- Total Bilirubin: 0.6 mg/dL
- Total Protein: 7.1 g/dL
- Albumin: 3.7 g/dL

Imaging

- Hgb: 11.4 g/dL
- WBC: 5.5 K/cu mm
- Plt: 202 K/cu mm
- CXR: clear lungs
- Mammogram: unremarkable
Problem Representation #1

61-year-old woman with history of tobacco use disorder, chronic lumbar back pain with radiculopathy, and breast cancer in remission presenting with cauda equina syndrome.
Prioritized Differential Diagnosis

- Chronic, progressive spinal stenosis
- Recurrent breast cancer with metastasis
- Disk herniation
- New malignancy
“Her degree of stenosis does not explain her incontinence symptoms. Other causes should be investigated.”
“Would you tell me, please, which way I ought to go from here?”
Problem Representation #2

61-year-old woman presenting with acute onset of cauda equina syndrome with progressive, spontaneous symptom improvement over 6 weeks.
Differential Diagnosis

Compressive
• Disk herniation
• Fracture
• Neoplasms
• Iatrogenic injury
• Chronic lumbosacral spinal stenosis
• Infection

Non-compressive
• Ischemia
• Infiltration
• Inflammation
• Infection

Tarulli AW. Disorders of the Cauda Equina. Continuum 2015; 21(1): 146-158
Differential Diagnosis

Compressive

• Disk herniation
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Non-compressive

• Ischemia
• Infiltration
• Inflammation
• Infection

Tarulli AW. Disorders of the Cauda Equina. Continuum 2015; 21(1): 146-158
Key Features

• Acute onset
• Self-limited
• Non-compressible
Differential Diagnosis

Non-compressive

• Ischemia
• Infiltration
• Inflammation
• Infection
Differential Diagnosis

Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection

- Dural AV Fistula
- Vascular Accident

Differential Diagnosis

Non-compressive

• Ischemia
• Infiltration • Sarcoidosis
• Inflammation
• Infection

Differential Diagnosis

Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection

  - Guillain-Barré
  - Vasculitis

Goodman BP. Disorders of the Cauda Equina. Continuum 2018; 24 (2, spinal cord disorders):584-602
Differential Diagnosis

Non-compressive

- Ischemia
- Infiltration
- Inflammation
- Infection

- Neurosyphilis
- Tuberculosis
- CMV + AIDS
- VZV
- HSV
HSV
61-year old woman with history of:

- CKDIII
- Inflammatory breast cancer (2007) in remission
- Herpes genitalis
- Chronic lumbar back pain
- Tobacco use disorder
Outside Records – Urgent Care Visit

- Chief complaint: vaginal and pelvic pain
- HPI: onset 1 week prior to cauda equina syndrome
- GU Exam: multiple, bilateral ulcerations of the labia and surrounding perineum
- Vaginal Swab: + Herpes Simplex Virus
Problem Representation #3

61-year-old woman presenting with acute, primary genital herpes infection, followed days later by acute onset of cauda equina syndrome, with progressive symptom improvement over 6 weeks.
Elsberg Syndrome

An infectious syndrome consisting of acute or subacute lumbosacral radiculitis, with or without myelitis, provoked by primary or reactivated herpes simplex infection.

Elsberg Syndrome – Diagnostic Criteria

• Clinical signs/symptoms of cauda equina syndrome

Elsberg Syndrome – Diagnostic Criteria

- Acute/subacute onset; no relapse
- Coexisting or recently preceding HSV infection

Elsberg Syndrome – Diagnostic Criteria

- MRI evidence of cauda equina involvement

Elsberg Syndrome – Diagnostic Criteria

- Clinical or MRI evidence of myelitis
- CSF pleocytosis
- HSV infection from CSF by PCR, culture, or IgM

Elsberg Syndrome – Natural History

- Neurologic clinical spectrum of HSV genital infections:
  - urinary retention (seen in 5-15% of primary infections)
  - aseptic meningitis
  - cauda equina syndrome
  - myelitis

- Self-limited, but may leave some degree of permanent neurologic impairment

- Treatment: in acute period, IV acyclovir +/- steroids

Follow Up

- History: continued symptom improvement
- Labs: negative HIV and syphilis
- Plan:
  - Offered consultation with neurology and additional diagnostics – she declined
  - Monitored for new neurologic symptoms – none to date
  - Suppressive acyclovir
“Tut, tut, child!” said the Duchess. “Everything’s got a moral, if only you can find it.”
Moral of the Story

- Not all causes of cauda equina syndrome are surgical emergencies (though we should proceed as if they are)
- non-compressive etiologies include ischemia, infection, inflammation, and infiltration
Moral of the Story

• Not all causes of cauda equina syndrome are surgical emergencies (though we should proceed as if they are)
  • non-compressive etiologies include ischemia, infection, inflammation, and infiltration

• Elsberg Syndrome is an uncommon manifestation of a common infection
  • Clinical spectrum of HSV genital infection includes radiculitis and myelitis
  • Diagnosis may be aided by laboratory evidence of HSV infection, CSF studies, and MRI
“Begin at the beginning,” the king said, very gravely, “and go on till you come to the end: then stop.”
References

- Goodman BP. Disorders of the Cauda Equina. Continuum 2018; 24 (2, spinal cord disorders):584-602
- Tarulli AW. Disorders of the Cauda Equina. Continuum 2015; 21(1): 146-158
Elsberg Syndrome - Epidemiology

- Most commonly caused by HSV-2
- Most often seen in sexually active young women
- Under-recognized; data limited to case reports and series
