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# A 5-Year-Old With Fever, Headache, Neck Stiffness, and Leg Pain

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A 5-year-old boy presented with fever, headache, fatigue, neck stiffness, and 2 episodes of nocturnal urinary incontinence, prompting a visit to the emergency department. He had experienced intermittent frontal headaches and leg and buttock pain for several months, which had worsened over the previous 2 weeks. His history was notable for a spinal hemangioma with vascular tract, but he was otherwise healthy. On examination, he was febrile and tachycardic. He held his neck slightly rotated to the right with limited range of motion in all directions due to pain. No focal neurologic deficits were noted, and sensation and deep tendon reflexes were intact bilaterally. He was able to bear weight on both legs. There was no spinal tenderness or limitation in range of motion of his back and hips. There were no cutaneous manifestations, including no sacral dimple. A complete blood count with differential revealed leukocytosis of  $31.98 \times 10^3/\mu\text{L}$  (78.6% neutrophils, 16% bands). C-reactive protein was elevated at 2.4 mg/dL (0–1 mg/dL), and serum electrolytes, liver function tests, uric acid, and lactate dehydrogenase were within normal limits for age. Blood cultures were obtained before admission. Here we present his case, diagnostic evaluation, ultimate diagnosis, and complications.

## CASE HISTORY WITH SUBSPECIALTY INPUT

### Drs Solano and Winningham (Pediatrics, Residents):

A 5-year-old boy was admitted to the general pediatrics inpatient service with fever, headache, fatigue, neck stiffness, buttock pain, and bilateral leg pain. During the 2 weeks before admission, he was less active than usual and complained daily of leg and buttock pain. His leg and buttock pain limited his ability to walk on some occasions. The patient's mother noted that he had been complaining of frontal headaches and buttock pain intermittently over the past 2 to 3 months, which was unusual for him, and that these symptoms had increased in frequency and severity over the previous 2 weeks. The day before admission, he was noted to

have a headache associated with a temperature of 100.8°F (38.2°C). He had a reported fever at home 2 weeks before admission, but it resolved within 24 hours and was associated with congestion. The patient's mother did not report any other fevers during his illness. Additionally, he had 2 episodes of nocturnal enuresis within the few weeks before presentation, which was new for him. On the day of admission, the patient began holding his head in a fixed position and could not look from side to side due to pain.

Dr Barnett, what is the differential diagnosis at this point?

### Dr Barnett (Pediatric Emergency Medicine):

Meningitis must be considered in any patient presenting with headache, fever, and neck stiffness.<sup>1</sup> The course of this patient's illness was

## abstract

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unusually intermittent and more indolent than one would expect with acute meningitis.<sup>1</sup> The additional complaints of leg pain and pain in the gluteal region led providers to broaden the differential diagnosis. The combination of this time course and history of symptoms involving both the lower extremities as well as the neck are concerning for an intraspinal or paraspinal abscess, vertebral osteomyelitis, discitis, sacroiliitis, or tumor, all with involvement of the meninges causing symptoms of meningitis. Trauma is less likely given the presence of fever and no history of repeated neck, back, or leg injuries.

#### **Drs Solano and Winningham:**

The patient had scabies 5 months before presentation but had an otherwise unremarkable past medical history per the initial report. After a more thorough review of his chart, it was discovered that the patient had a cutaneous hemangioma overlying the spine at birth. An ultrasound performed at 4 months of age revealed a thin curvilinear vascular track extending from the superficial posterior lumbosacral cutaneous region anteroinferiorly to the region of the deep fascial layer. No follow-up occurred. He had been taking ibuprofen and acetaminophen intermittently over the past few months for headaches as well as for fever on the day of admission. He did not take any daily medications.

On presentation to the emergency department, he had a temperature of 101.8°F (38.8°C) and a heart rate of 157 beats per minute. His respiratory rate was 28 breaths per minute, and his blood pressure was 120/68 mm Hg. His oxygen saturations were recorded at 97%. He was ill appearing, and his movements were restricted by pain, primarily in his neck. It was a severe, dull pain that did not radiate. It worsened with movement and stabilized with rest, but he could not find a position to

give him complete relief. He held his neck slightly rotated to the right and moved his whole body with neck movement. His extraocular muscles were intact, and his pupils were equal and reactive. His pharynx was nonerythematous, but his mucus membranes were moderately dry. Aside from tachycardia, his heart, lung, and abdominal exams were normal. All parts of his spine were nontender to palpation and without abnormal cutaneous findings. No focal neurologic deficit was noted, and he had a normal motor examination. The perianal region was inspected and was normal.

Dr Barnett, what are the benefits to performing imaging before any other procedure or treatment based on the information that was known at this point?

#### **Dr Barnett:**

Fortunately, the patient was clinically stable and neurologically intact, so no intervention was emergently required. Excluding paraspinal abscess or tumor is essential before an invasive procedure involving the spine. Given the time course of his illness and clinical state, it seemed prudent to wait on treatment with antibiotics until a more definitive diagnosis was reached.<sup>1</sup> Ultrasound was not considered because there were no abnormal findings on skin examination. MRI of his brain and spinal cord was pursued.

#### **Drs Solano and Winningham:**

In the emergency department, laboratory results were as follows: white blood cell (WBC) count  $31.98 \times 10^3/\mu\text{L}$  (78.6% neutrophils, 16% bands), hemoglobin 12.8 g/dL (11.5–13.5gm/dL), platelet count  $406 \times 10^3/\mu\text{L}$ , erythrocyte sedimentation rate 8 mm/hour (0–13 mm/hour), C-reactive protein (CRP) 2.4 mg/dL (0–1 mg/dL), and creatinine kinase 34 unit/L (75–230 unit/L). Serum electrolytes, liver function tests, uric acid, and lactate dehydrogenase were

within normal limits for age. Blood cultures were obtained and remained negative. Chest radiograph was normal. MRI of the brain and total spine was performed.

Dr Zinkus, what features on the patient's imaging are important for the clinician to recognize?

#### **Dr Zinkus (Pediatric Neuroradiologist):**

The MRI of the patient's brain revealed no intracranial pathology. However, the MRI of his spine demonstrated an intraspinal mass, which filled the distal spinal canal and produced ventral displacement of the nerve roots of the cauda equina (Fig 1). There was enhancement of the nerve roots of the cauda equina compatible with inflammation. The mass appeared intradural with expansion of the distal thecal sac. The mass was heterogeneously hyperintense on T2, hypointense on T1, and contained peripheral contrast enhancement. Because no internal or solid-appearing contrast enhancement was present, the mass was most likely cystic given the appearance on T1 and T2. In addition, the mass demonstrated mildly reduced diffusion on diffusion-weighted imaging. Diffusion signal abnormalities are nonspecific but may be seen with purulent material or highly cellular tumors in this context. Given the presence of reduced diffusion, the 3 primary differential considerations would include intraspinal abscess, cellular tumor, or intraspinal dermoid/epidermoid. In this case, however, T2-weighted imaging also demonstrated a sinus tract that extended between the L4–5 spinous processes and into the midline aspect of the spinal canal. Given the presence of a sinus tract, the main differential considerations for this mass could be narrowed down to intraspinal abscess and intraspinal dermoid/epidermoid with or without secondary infection.



**FIGURE 1**  
The sagittal cut of the lower thoracolumbar spine and sacrum shows an intraspinal mass consistent with an abscess, which fills the distal thecal sac (solid arrow). There is an associated sinus tract that extends from the subcutaneous tissues into the spinal canal at the level of the mass (dashed arrow).

**Drs Solano and Winningham:**

In the emergency department, vancomycin and ceftriaxone were started for broad coverage to treat the suspected intraspinal abscess noted on the MRI. He received a total of 40 mL/kg of bolus isotonic fluid followed by the initiation of maintenance intravenous (IV) fluids. Acetaminophen and ibuprofen were given for pain management, and he was admitted to a general pediatrics team with a plan for operative debridement the next morning.

Dr Igbaseimokumo, what surgical interventions are considered once an intraspinal abscess is diagnosed? How urgently must surgery be performed?

**Dr Igbaseimokumo (Pediatric Neurosurgery):**

The surgical approach to a spinal abscess is guided by the presence or absence of mass effect from the collection. The presence of cauda equina syndrome (loss of bladder and bowel control, with or without leg weakness), compressive myelopathy or radiculopathy, or intolerable pain in the face of a sizeable collection generally indicates the need for

urgent surgical decompression rather than antibiotic therapy only.

For this patient, given the large size of the lumbar abscess with compression of the nerve roots seen on MRI, surgical drainage of the abscess was prudent. This was achieved by a laminectomy and surgical dissection of the nerve roots and evacuation of the abscess. The lower part of the abscess was densely matted together with the nerve roots, and there was minimal pus collection within. Thus, it was considered unsafe and unnecessary to try to tease the nerve roots apart to wash out the semisolid granulation mass that constituted the abscess at this level.

The surgical path is mostly guided by whether the abscess is anterior or posterior to the spinal cord as well as the vertebral level of the abscess: that is, whether it is in the cervical, thoracic, or lumbar spine. The timing of surgery, whether it is emergent (immediately) or urgent (within 24 hours), is determined by the severity of the symptoms, especially the presence or absence of progressive neurologic deficit such as complete loss of bladder or bowel control, which this patient fortunately did not have. The extent of surgery is tailored to decompress the nerve roots and spinal cord. Reconstruction of collapsed vertebrae in the presence of a spinal abscess is beyond the scope of this discussion and fortunately not common.

**Drs Solano and Winningham:**

On day 2 of admission, neurosurgery performed a laminectomy, abscess incision and drainage, and resection of the sinus tract. Cerebral spinal fluid (CSF) was collected during the procedure revealing a white blood cell count of 3825 white blood cell/ $\mu$ L with 79% segmented neutrophils, glucose <20 mg/dL, protein 332 mg/dL, and red blood cells 286/ $\mu$ L. Gram stain was performed and revealed many Gram-positive cocci in clusters,

few white blood cells, and few epithelial cells.

Pathology revealed a dermoid cyst with an associated sinus tract remnant. CSF and wound cultures grew *Staphylococcus epidermidis*, which was susceptible to oxacillin and rifampin. His therapy was switched to oxacillin and rifampin on day 3 of admission.

Drs Myers and Al Zubeidi, what are the most likely organisms identified in patients with an infected dermoid cyst associated with a dermal sinus tract? What is the most appropriate treatment regimen?

**Drs Myers and Al Zubeidi (Pediatric Infectious Disease):**

Empirical antibiotic choice should target the most common infectious pathogens, specifically skin flora, because infection in children is mostly secondary to direct contiguous spread from a preexisting congenital defect in the spinal cord. In contrast, these infections arise from hematogenous spread in adults. In a review of 29 cases of pediatric intradural spinal abscesses, *Staphylococcus* spp, *Streptococcus* spp, enteric Gram-negative bacilli, and anaerobes caused the majority of cases in which an isolated organism was identified highlighting the importance of both broad coverage and obtaining routine aerobic and anaerobic cultures.<sup>2</sup> Intraspinal abscesses caused by *S epidermidis* are well described in association with a congenital dermal sinus tract.<sup>3</sup>

*Mycobacterium tuberculosis* has been rarely reported and when present usually presents in patients with antecedent history of tuberculosis meningitis.<sup>2,4,5</sup> In children with recurrent urinary tract infections, one should consider Gram-negative bacteria, especially *Escherichia coli*, as a causative pathogen. Others such as *Pseudomonas* (in injection drug users), anaerobic bacteria (eg, *Actinomyces* spp), *Nocardia* spp, *Mycobacteria* spp,



fungi, and parasites have been reported.<sup>6</sup> The fact that this patient's symptoms were prolonged with more of a chronic presentation makes *S epidermidis* a more likely pathogen because coagulase-negative staphylococci tend to cause more indolent infections without high spiking fevers and evolve over weeks to months. If the presentation had been acute in nature (eg, symptoms for a few days to a week) *Staphylococcus aureus* or an enteric Gram-negative organism would be a more likely pathogen. Thus, obtaining the patient history in terms of length of symptoms as well as symptom severity helps to tailor the differential diagnosis to the most likely pathogens. Pending surgical intervention and culture results, empirical antibiotic therapy should provide broad-spectrum coverage. A third- or a fourth-generation cephalosporin in addition to vancomycin is the preferred option. Intravenous therapy should continue for a minimum of 4 to 6 weeks.<sup>7</sup>

#### **Drs Solano and Winningham:**

Electrolytes were serially monitored, and the patient's serum sodium level was noted to be 132 mmol/L (135–145 mmol/L) on postoperative day 2. Fluids were initially restricted, but a repeat sodium level drawn several hours later was 129 mmol/L (135–145 mmol/L). Urine sodium was above normal suggesting a cerebral salt wasting process. He was transferred to the Pediatric ICU for closer neurologic monitoring.

Dr Benton, what is the reason for his hyponatremia, and how is it related to his underlying disease process?

#### **Dr Benton (Pediatric Intensivist):**

The common 2 disorders associated with hyponatremia are syndrome of inappropriate antidiuretic hormone (SIADH) and cerebral salt wasting (CSW). Disorders of sodium homeostasis are common with central nervous system (CNS)

injury. It is important to be able to distinguish between the 2 disorders because the treatments are different.<sup>8</sup> The key difference between CSW and SIADH is volume status of the patient. CSW is characterized by decreased total body water, whereas SIADH is characterized by increased total body water. Typically, patients with CSW will have normal or elevated urine output in contrast to SIADH, which typically is associated with decreased urine output.<sup>8</sup> All other laboratory markers (low plasma osmolality, high urine osmolality, hyponatremia) are similar. This patient's hyponatremia was most consistent with CSW and was most likely related to meningitis rather than the epidural abscess itself.<sup>9</sup> The exact pathophysiology of CSW is unknown, but the common end point of all theories is renal wasting of sodium. One theory suggests that there is a disruption in the sympathetic nervous system, which disrupts the renin-aldosterone axis leading to impaired sodium reabsorption in the proximal tubule. Another theory is that brain natriuretic peptide is released in response to the CNS injury which decreases sodium reabsorption in the kidney.<sup>10</sup> The salt wasting results in water loss. Treatment of CSW focuses on rehydration and salt repletion. This disorder is typically transient.

#### **FINAL DIAGNOSIS AND DISCUSSION: INTRADURAL SPINAL ABSCESS**

##### **Drs Myers and Al Zubeidi:**

Intradural spinal abscesses are uncommon in both children and adults. Hart reported the first case more than a century ago in 1830, and only 100 cases have been reported since, the majority in adults.<sup>11</sup> In adults, hematogenous seeding may occur from diabetes-associated infections, complicated urinary tract infections, surgical site infections, cardiac infections including bacterial endocarditis, bronchiectasis-associated infections, or, less

frequently, infections due to spinal abnormalities.<sup>12</sup>

In contrast, most pediatric cases are caused by contiguous spread from the skin from an underlying congenital spinal defect.<sup>13,14</sup> In a recent review, more than half of the cases had an underlying dermal sinus tract, and similar to this patient's case, a third of these cases had defects identified before presentation. The majority of children with intradural spinal abscesses present before 5 years of age (range 2 weeks–17 years of age). The clinical presentation varies depending on age, location of the abscess, duration of symptoms, and size and number of abscesses.<sup>15</sup> This patient's clinical presentation with fever, headache, back pain, gait abnormality, bladder dysfunction, and weakness are typical symptoms of patients with spinal intradural abscesses; although, in some cases, cauda equina syndrome is more prominent.<sup>13</sup> Spinal cord injury is caused by compression from the abscess, but ischemia can also develop due to vascular occlusion, which may occur in the setting of septic thrombophlebitis.<sup>6</sup> This patient's additional symptoms of neck pain and stiffness suggest the infection had spread to his CSF, causing meningitis. The additional history of spinal dermal sinus tract in this patient supported this clinical suspicion and prompted more in depth evaluation.

Laboratory abnormalities are usually nonspecific. The white blood cell count is elevated in about two-thirds of patients, and C-reactive protein and erythrocyte sedimentation rate are elevated in most patients. When CSF is examined, pleocytosis and elevated protein levels are often seen due to parameningeal irritation. Radiologic imaging with MRI is able to provide detail with regard to size and location of the abscess, as well as involvement of surrounding structures.<sup>6</sup>

The mainstay of therapy is pain relief and surgical debridement of infected tissues with laminectomy combined with IV antibiotics, which are initially broad spectrum to provide coverage of *Staphylococcus* spp and Gram-negative bacilli. Definitive therapy is guided by culture and susceptibility results. Steroids may be used to decrease spinal cord compression and neurologic deterioration in patients awaiting surgical intervention, but there have been some reports of worse outcomes with steroid use. Outcome is variable and depends on duration of symptoms and degree of neurologic deficits before surgical decompression.<sup>16</sup>

### Drs Solano and Dr Winningham:

With enteral NaCl supplementation, the patient's sodium levels normalized. He was transferred from the ICU to the general pediatrics service on postoperative day 3. He continued to have fevers, headache, vomiting, and back pain, prompting repeat imaging, which showed meningeal enhancement. By postoperative day 6, he defervesced, and his pain was well controlled. His activity level increased, and his oral intake improved to baseline. He was discharged after 17 days of inpatient IV antibiotic therapy. A peripherally inserted central catheter was placed to allow for continued IV therapy at home. After 29 days of therapy, he had follow-up evaluation in the infectious diseases clinic. The peripherally inserted central catheter line was removed at that visit. The patient followed up in neurosurgery clinic 2 months after hospital discharge. Repeat MRI at that time showed no new fluid collections.

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### ABBREVIATIONS

CSF: cerebral spinal fluid  
 CSW: cerebral salt wasting  
 IV: intravenous  
 SIADH: syndrome of inappropriate antidiuretic hormone

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