Case Report

Sacral Tumor

Urgent message: Each case—and patient presentation—in urgent care must be evaluated on its own merits. Rare diagnoses are possible and "benign" back pain complaints sometimes are not.

HEATHER VARLEY, PA-C, and WILLIAM GLUCKMAN, DO, MBA, FACEP

Introduction

Each day in the urgent care setting, we are presented with a range of various pain complaints, from headaches, to back pain, to extremity pain. These complaints may be the result of injury, overuse, infection, arthropathy, or have no clear underlying cause. Although many of the cases we encounter in the outpatient clinic setting are benign and will respond to initial conservative management, it is important not to group all pain complaints together with a similar clinical work up and treatment approach. Each case must be addressed individually, with a careful history and thorough physical exam. Serious causes for pain must be ruled out, and other causes for pain appropriately identified and managed.

The following example is a case in which the patient had few risk factors but presented with complaints and a physical exam concerning for a more serious cause for back pain. This case highlights the importance of looking at the whole picture, and keeping rare and potentially dangerous diagnoses in your differential amongst the sea of vague pain complaints.

Case Presentation

A 30-year-old otherwise healthy female who was newly married and had no children presented to the urgent care center after a slip in the shower on a shampoo bottle that morning. She fell backward and landed on her lower back and buttocks, right elbow, and the back of her head. The patient presented complaining primarily of moderate to severe pain of the coccygeal region,

Heather Varley is a full-time Physician Assistant and **William Gluckman** is President & CEO of FastER Urgent Care in Morris Plains, NJ.

8/10 in intensity, non-radiating. She denied loss of consciousness, current headache, nausea, vomiting, visual changes, numbness, tingling, lower extremity weakness, saddle anesthesia, bowel or bladder incontinence or retention. The elbow pain had resolved, and physical exam revealed no ecchymosis, swelling or limitation of movement. All other exams were unremarkable aside from localized tenderness over the sacrum and coccyx, without crepitus or deformity.

An x-ray was performed of the lumbosacral spine and coccyx, and read as normal. No apparent fracture was detected by the urgent care provider or the over-reading radiologist. The patient was discharged on non-steroidal anti-inflammatory drugs (NSAIDs), with oxycodone/acetaminophen for pain, and was instructed to apply ice to the affected area. She was feeling better

upon follow-up call a few days later.

The patient returned 3 weeks later complaining of 4 days of a return of the pain in her lower back and coccyx. She described the pain as a sharp ache that was 9/10 in intensity and increased at night, specifically when lying down. She noted numbness to her hips, perineum, and anterior proximal thighs. The patient also noted constipation, which she attributed to her use of pain medication over the past few days. She also reported mild urinary retention and a sensation that she had to urinate but was unable to intermittently over the past 4 days. She was able to walk and range her back without issue. She denied fever, chills, headache, dizziness, weakness, abdominal pain, nausea, vomiting, and dysuria. Her last menstrual period was 2 weeks prior and normal. After further questioning, patient did note that she had been experiencing intermittent lower back pain at night for the past year.

Observation/Findings

Evaluation of the patient showed the following:

- T: 97.2
- RR: 16
- P: 71
- BP: 121/77
- O2: 99% RA
- Weight: 110 lb Height: 5'3"

Physical exam revealed a well-appearing female in no acute distress. Positive exam findings included: *Right Anterior thigh (dermatome L1-S3)*—light touch sensation diminished, sharp/dull sensation diminished, 2-point discrimination intact, pain sensation intact, vibration sense intact.

Left anterior thigh (dermatome L1-S3)—light touch sensation diminished, sharp/dull sensation absent, 2-point discrimination intact, pain sensation diminished, vibration sense intact.

Right and left medial leg (dermatome L4)—light touch sensation diminished, sharp/dull sensation intact, 2-point discrimination intact, pain and vibration sense intact.

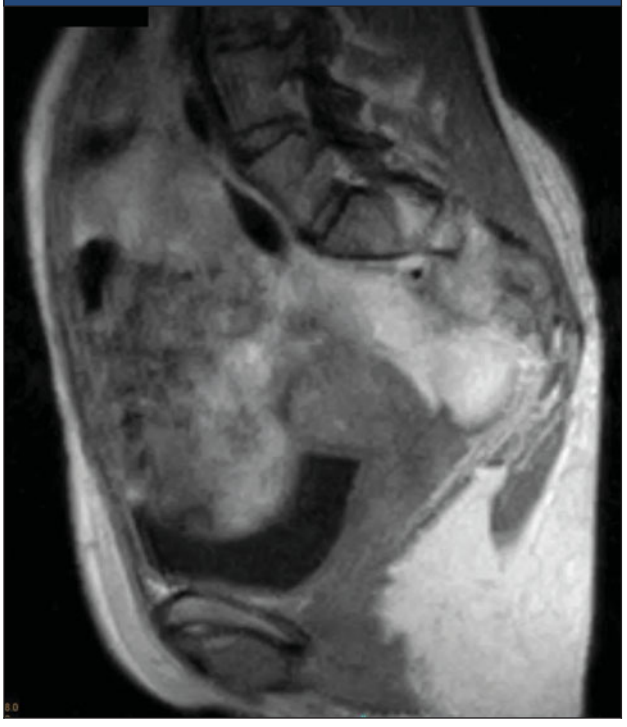
Lateral leg/medial foot (L5 dermatome)—sensation intact throughout to light touch, sharp/dull, 2-point discrimination, pain and vibration.

Lateral leg/dorsal foot (L5 dermatome)—sensation intact throughout to light touch, sharp/dull, 2-point discrimination, pain and vibration.

Lateral ventral foot (S1 dermatome)—sensation intact throughout to light touch, sharp/dull, 2-point discrimination, pain and vibration.

Proprioception was intact.

Figure 1. MRI of the sacrum and coccyx



Overall it was noted that decreased sensation was greater in the left thigh and pelvic region compared to the right thigh and pelvic region.

Cranial nerves II-XII and all cerebellar tests were within normal limits. Strength was 5/5 and equal bilaterally in both the upper and lower extremities. Reflexes and pulses were 2+ throughout. Gait was normal. Musculoskelatal examination of the lower back was unremarkable, with no point tenderness and full range of motion. The abdomen was soft and non-tender with normal bowel sounds in all four quadrants.

Diagnostic Studies

Because of concern about nerve impingement/cauda equina syndrome, the patient was referred to a local imaging center for stat magnetic resonance imaging (MRI). MRI without contrast of the lumbosacral spine was obtained, with the presumed diagnosis of lumbar disc displacement with myelopathy vs. lumbar disc rupture with myelopathy. The patient was prescribed oxycontin, 10 mg Q12 with Nucynta, 75 mg 1-2 tabs q4-6 hrs PRN for breakthrough pain.

Diagnosis

Primary sacral bone tumor

MRI of the lumbar spine demonstrated no acute lumbar disc herniation/extrusion or acute osseous trauma to the lumbar spine. MRI of the sacrum and coccyx demonstrated a large multiloculated expansile cystic lesion centered on the mid sacrum with pathologic fracture (**Figure 1**). Findings were thought to represent a large aneurysmal bone cyst or giant cell tumor of the sacrum.

In order to further evaluate the mass and better define the osseous anatomy, an MRI with contrast and computed tomopgrahy (CT) without contrast of the sacrum and coccyx were ordered.

The patient was referred to Memorial Sloan-Kettering Cancer Center in New York City, where she was admitted for further work up and pain management, even though a tissue diagnosis had not yet been obtained.

The patient underwent CT scanning and a biopsy, which revealed chondroid chordoma of the sacrum.

Course and Treatment

The patient was placed on steroids in order to decrease inflammation and impingement on spinal nerves. She underwent a radical surgical resection of the tumor, with removal of her sacrum and coccyx beginning at the level of S2. Two lesions in the right and left pelvis were also removed through interventional radiology procedures. Chemotherapy and radiation had no benefit in the management of her chondroid chordoma, and were therefore not recommended as part of the treatment course. She remained in the hospital for 8 weeks for pain management, occupational therapy, physical therapy, and bowel and bladder training. She was left with residual bowel and bladder incontinence. She self-catheterizes her bladder 3 hours and is on a bowel regimen, taking Metamucil at night and a suppository in the morning in order to have bowel movements. She does not require a colostomy at this time. She remains in physical therapy three times per week for lower back and lower extremity strengthening and is now ambulating without assistance. She remains on methadone, 10 mg TID and Neurontin TID for pain management. The patient has frequent positron emission tomography and CT scans in order to monitor for recurrence and metastases, which to date, have remained clear.

Discussion

Chordomas are part of a family of cancers called sarcomas, which include cancers derived from the cells of bones, cartilage, muscles and other connective tissue. Chordomas most closely resemble cartilage in their histologic tissue make up. Chordomas arise from cellular remnants of the notochord, the cells in the embryo that form the foundation for development of the spinal cord. The notochord regresses during fetal life as the spinal cord develops, however, some notochord cells do normally remain after birth embedded in the clivus of the skull and the sacrococcygeal regions of the spine. Rarely these remaining notochord cells go through a malignant transformation that leads to formation of a chordoma. Because of this local transformation, chordomas are most commonly found along the neuraxis, intracranially at the clivus, or in the sacrum at the base of the spine. Chordomas are rare slow-growing malignant bone tumors, accounting for 1% to 4% of primary bone tumors. However, of the primary bone tumors affecting the sacrum, more than half are chordomas. In the United States the annual incidence of chordoma is 1 in 1 million, with 300 new cases diagnosed each year. Because chordomas are typically located in close proximity to crucial structures such as the spinal cord, brain stem, and important nerves and arteries, they can be very difficult to treat, requiring highly specialized surgical and oncologic care, with close long-term follow-up.

Chordomas can occur at any age, but they are more common in adults, with a median age at diagnosis of 49 for skull base chordomas and 69 for sacral chordomas. The ratio of males to females affected by this rare tumor is approximately 1.6 to 1. There have been no definite identified causes for chordoma, however, some studies have indicated a possible genetic predisposition for development of the disease.

The symptoms of chordomas depend on their location. Patients with clival chordomas at the base of the skull typically present with headache, facial pain or paralysis, double vision, changes in hearing or tinnitus, difficulty swallowing, or hoarse voice. Sacral chordomas often do not cause symptoms until they are very large and patients with them may present with back pain, lower extremity pain, lower extremity weakness, numbness or tingling, rectal dysfunction, urinary retention or incontinence, erectile dysfunction, or in some cases, a palpable sacral mass.

Diagnosis of chordoma typical involves neurologic examination, imaging studies including MRI and/or CT of the affected area, followed by guided tissue biopsy. The three histologic types of chordoma are classical ("conventional"), chondroid, and dedifferentiated.

Chondroid chordomas tend to have a more indolent clinical course and be less aggressive overall than classical chordomas. Dedifferentiated chordomas are typically more aggressive and more likely to metastasize.

Surgical resection is the mainstay of treatment. Complete surgical removal of the tumor while preserving vital structures is essential in order to offer the best chance of survival and decrease the possibility of localized recurrence and metastases. Recent advances in surgical, imaging, and interventional radiology techniques have allowed surgeons to perform complete tumor excisions more commonly and successfully. High-dose radiation typically is required for successful treatment of chordomas. However, the proximity of vital neurologic structures to chordomas limits the dose of radiation that can be safely utilized. Therefore, radiation tends to be reserved for cases in which the entire tumor cannot be safely excised, or to shrink the tumor prior to resection in order to avoid damage to critical structures. Even with advanced techniques, complete resection of chordomas often comes at the expense of certain neurologic structures, impacting long-term function and quality of life in these patients. For example when resecting a sacral tumor, both S3 nerves must be preserved in order to maintain urinary and bowel function. If both S3 nerves are resected then patients will likely need to intermittently self-catheterize and use bowel medications. If both S2 nerves are resected, the result is complete urinary and bowel incontinence, requiring consistent urinary catheterization and a bowel regimen, as was the case with the patient in our case analysis. There are no chemotherapy drugs currently approved for treatment of chordoma, but recent trials have demonstrated a moderate response to the PDGFR inhibitor imatinib.

Prognosis for chordoma is different for each individual case. Factors such as patient age, tumor size and location, histological subtype, and extent of resection all affect clinical outcomes.

Take-Home Point

When dealing with a patient who presents with back pain, it is essential to identify possible risk factors for potentially serious spinal conditions and to ask all of the "red flag" questions to every single patient. A careful history including the patient's age, cancer history, immunosuppression, drug use, changes in weight, fever, chills, prolonged steroid use, urinary symptoms, recurrent UTI, prolonged pain or pain not improved with rest, may lead you to consider cancer or infection. A history of trauma or osteoporosis may raise suspicion for spinal fracture. Sudden onset of urinary retention or overflow incontinence, fecal incontinence or decreased rectal tone, constipation, saddle anesthesia, weakness in the lower extremities , or sexual dysfunction are concerning for cauda equina syndrome or severe neurologic compromise, and require an emergent work-up.

The patient in this case study came in believing her pain was associated with a fall 3 weeks prior and attributed some of her abnormal bowel and bladder function to the medication she was using to manage that pain. At first glance, you might agree with the patient and attribute her pain to the recent fall and the constipation to the opiates, but after questioning and physical exam, it was clear that she had serious neurologic clinical findings suggestive of spinal cord compression, and that she would require an immediate referral with an extensive work-up in order to offer the best prognosis. We had the luxury of being able to obtain an immediate MRI, although transfer to an emergency room would have been appropriate. ■

Bibliography

- Cheng EY, Ozerdemoglu RA, Transfeldt EE, Thompson RC. Lumbosacral chordoma: prognostic factors and treatment. *Spine.* 1999; 24(16): 1639.
- Chordoma. Mount Sinai Hospital website. Available at: http://www.mountsinai.org/patient-care/health-library/diseases-and-conditions/chordoma. Accessed May 1, 2013.
- Chordoma. Mayo Clinic website. Available at: http://www.mayoclinic.org/chordoma/. Accessed May 15, 2013.
- Gardner P, Snyderman C, Malyapa R. Understanding chordoma. Chordoma Foundation Website. Available at: http://www.chordomafoundation.org/understanding-chordoma/. Accessed May 15, 2013.
- Ishii K, Chiba K, Watanabe M, et. al. Local recurrence after S2-3 sacrectomy in sacral chordoma: report of four cases. *Journal of Neurosurgery: Spine.* 2002; 97(1): 98-101.
- Sciubba DM, Cheng JJ. Petteys RJ, et. al. Chordoma of the sacrum and vertebral bodies. *The Journal of the American Academy of Orthopaedic Surgeons*, 2009; 17(11) 708-717.
- Walcott BP, Nahed BV, Mohyeldin A, et. al. Chordoma: current concepts, management, and future directions. *The Lancet Oncology*, 2012; 13(2) 69-76.