CLINICAL VIGNETTE

Back Pain due to Retroperitoneal Liposarcoma

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Case Presentation

A 66-year-old man with type 2 diabetes mellitus and chronic kidney disease presented to primary care clinic with intermittent nausea, anorexia, and right-sided low back pain that started three to four months prior. It had gradually worsened over the past two months. He particularly noted pain with ambulation, which lessened with lying or sitting, but would not go away completely. He described the discomfort as an aching pain deep in the lower back from the middle to the right side with no radiation down the legs. He denied any recent trauma to the back or heavy lifting. There was no saddle anesthesia, no dysuria or hematuria. While he would occasionally have urinary leakage if he did not make it to lavatory while taking his diuretics, he was generally continent of both urine and stool. Past medical history also includes morbid obesity, hypertension and non-ischemic cardiomyopathy.

Vitals were normal with the exception of a BMI of 43. He appeared uncomfortable but otherwise in no acute distress. On cardiac examination, he had a regular rate and rhythm with no murmurs, rubs, or gallops, although heart sounds were distant. He had 1-2+ lower extremity pitting edema to the mid shins bilaterally with moderate overlying redness, which was his baseline. His abdomen was protuberant with a large pannus and intra-abdominal structures could not be well palpated. His back exam revealed mild tenderness to palpation along the L2-3 spinous processes and also on the right paraspinal muscles at the same level. There was no tenderness to palpation along the sacroiliac joints. Flexion was limited to 30°. Strength was normal for both plantar and dorsiflexion. Gait was normal.

Lumbar X-ray revealed 1.6cm of anterolisthesis and moderately severe L5-to-S1 degenerative disc disease. He was diagnosed with mechanical low back pain and pursued conservative treatment with acetaminophen, local heat packs, baclofen, and lidocaine patches.

Two months later, he was admitted for decompensated congestive heart failure and developed right-sided abdominal pain and non-bloody watery diarrhea after discharge. He also had persistent chronic right flank pain. A complete blood count showed a leukocytosis with a neutrophil predominance and a chronic, normocytic anemia with a hemoglobin of 11.6mg/dL. Basic metabolic panel showed severe hyperglycemia with glucose 663 and a non-gap metabolic acidosis. His GFR decreased from a baseline of 40 to 32mL/min. CT of the abdomen revealed a large 20cm right-sided retroperitoneal mass encasing the right kidney and part of the inferior vena cava. He was admitted and underwent a CT-guided biopsy of the mass, which showed a well-differentiated lipomyosarcoma. His case was presented to the interdisciplinary tumor board for further management.

Epidemiology

Retroperitoneal liposarcomas are rare tumors that account for <1% of malignancies. In general, sarcomas tend to occur along the appendicular skeleton, and retroperitoneal liposarcomas account for 10-15% of total sarcomas.[1] However, most retroperitoneal masses are malignancies and one-third of those are sarcomas, of which liposarcomas are the most common. Men and women are equally affected with average age of occurrence in the 50s.

Clinical Presentation

This patient had a classic presentation as most are asymptomatic until the tumor enlarge to cause mass effect on surrounding organs. This causes pain, abdominal fullness, lower extremity edema, localized neurological and musculoskeletal symptoms, as well as gastrointestinal complaints such as early satiety. Once symptoms have developed, tumors are locally advanced and average approximately 15cm at the time of diagnosis.[2]

Evaluation and Management

Contrast-enhanced CT imaging of the abdomen and pelvis is the most helpful radiographic diagnostic tool. MRI can be a secondary option if there is contrast allergy, but motion artifact may reduce the accuracy of the image. PET scanning may identify distant metastases if histology indicates a higher grade lesion, but is generally unhelpful and not standard in the evaluation of a typical liposarcoma. If a high-grade subtype such as a leiomyosarcoma originating from smooth muscle is found instead of a typical liposarcoma which originates solely from stromal fat tissue, then CT scan of the thorax is also warranted since higher grade tumors tend to spread to the lungs. The liver is a secondary source for metastatic extension.

While the differential diagnosis for retroperitoneal masses includes lymphoma, primary germ cell tumors, metastatic testicular cancer, schwannomas, Castleman's disease, retroperitoneal fibrosis, and other retroperitoneal cancers such as pancreas, duodenum, kidneys, and adrenals. These tend to have different appearance on imaging. Liposarcomas tend have the same signal intensity as regular adipose tissue, although occasionally with septations or areas of hemorrhage.[3]

In addition to imaging, percutaneous biopsy is standard. While there have been concerns about seeding tumor into the surrounding tissue or triggering hematogenous spread during the biopsy, the risk is exceedingly low, less than 0.5%.[4]

Management is almost exclusively surgical resection. Resection is only avoided when there is extensive multi-vessel involvement such as aorta, inferior vena cava, and superior mesenteric artery. Distant metastases are exceedingly rare with well-differentiated liposarcomas, as are peritoneal implants, or extensive spinal cord involvement. There is often need to resect other organs such as the spleen, bowel, or kidney to achieve a complete resection, operative mortality can be high. When blood vessels are involved, there is a 4% operative mortality rate.[5]

In general, radiation and chemotherapy have not been the standard of care for retroperitoneal liposarcomas. The concern with radiation is the large treatment field which includes many other structures with a lower radiation tolerance. Some institutions may include preoperative/neoadjuvant radiation therapy on a case-by-case basis, particularly if radiation could shrink the tumor enough to allow for surgical resection. In addition, large tumors can shield other structures from radiation. Postoperative radiation has been tried, but is usually unsuccessful given the empty surgical bed and risk to surrounding organs.[6]

While resection plus chemotherapy is common in extremity sarcomas, retroperitoneal liposarcomas are generally chemotherapy-unresponsive. Trials have shown no improvement in overall mortality.

Prognosis

Unlike most other tumors, there is very little prognostic contribution from tumor size. The most important prognostic factors are histology—with a well-differentiated liposarcoma having the best prognosis—and completeness of the resection. Unfortunately, even with complete resection, there is a high rate of recurrence. Local-regional recurrence, is 50% at 5-years.[7] Five-year survival also is about 50% for individuals who have had some form of surgical resection, and up to 60% to 80% for tumors that were completely resected at the time of the primary surgery.[8] Debulking surgeries are not recommended for asymptomatic growth, but repeat resections are sometimes pursued for palliation.

Case Conclusion

The patient was not considered a candidate for adjuvant radiation or chemotherapy due to obesity and habitus which would have required excessively large radiation window. He was also not a chemotherapy candidate given his history of cardiomyopathy, plus the unlikely responsiveness of his tumor. He opted for an open right radical nephrectomy with tumor resection, which was potentially curative, but considered high risk given his comorbidities. He underwent resection with an interdisciplinary team of General, Vascular, and Urologic surgeons. They were able to successfully resect the tumor en bloc, but discovered a small tear in the inferior vena cava from a tumor side branch causing massive intraoperative hemorrhage.

The patient developed hypovolemic shock with pulseless-electrical activity cardiac arrest, and responded to CPR. He required replacement of 8 liters of fluids and 16 units of blood products, with control of bleeding. The patient also developed ventricular fibrillation and successfully defibrillated and regained stable rhythm.

He survived resuscitation, was extubated on the first postoperative day, without apparent neurological sequelae. His back pain also resolved. After discharge he underwent serial imaging to monitor his tumor. At 14 months post-op, a recurrence was detected with a large mass in the left retroperitoneal space. He was asymptomatic and closely monitored for 9 months, when he died from congestive heart failure.

Figures

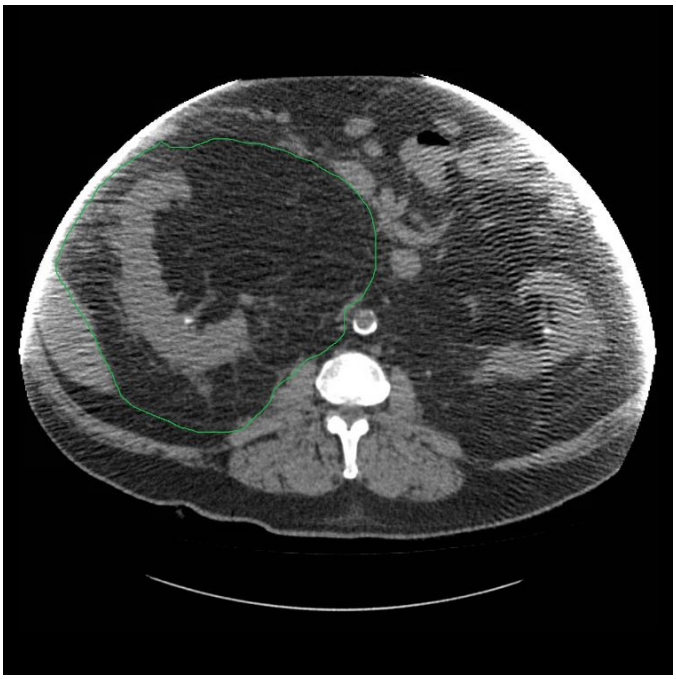

Figure A. Axial view of the liposarcoma around the time of diagnosis

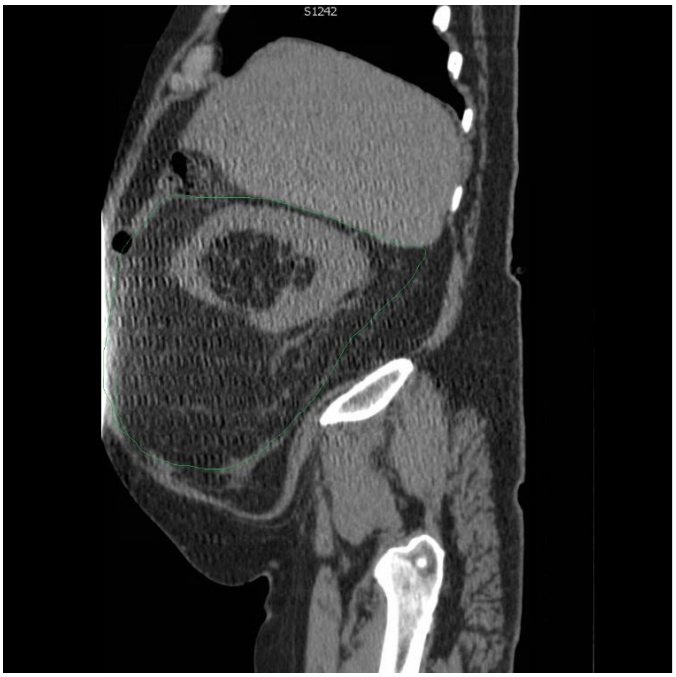

Figure B. Sagittal view of the liposarcoma around the time of diagnosis

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