



Surgical management of a large retroperitoneal liposarcoma: A case study

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ARTICLE INFO

Keywords:

Retroperitoneal sarcoma
Well-differentiated liposarcoma

ABSTRACT

The patient was a 45-year-old male who initially presented with a left hydrocele. During radiographic work-up, a 26 cm right retroperitoneal lipoma was incidentally discovered. Despite a recommendation for preoperative radiation therapy followed by surgery from the sarcoma multispecialty team, the patient opted for surgery alone, in the hopes of avoiding damage or loss of his right kidney. Following surgical excision of the 39 cm well-differentiated liposarcoma, with removal of the perinephric fat adjacent to the tumor thereby preserving the kidney, he was discharged home after two nights in the hospital. Follow-up imaging eight months later showed no recurrence.

Introduction

Retroperitoneal sarcomas are generally rare and comprise a highly heterogeneous group of mesenchymal neoplasms that are often large and locally invasive at presentation.^{1,2} The size of the retroperitoneal sarcomas is a significant predictor of overall survival, local recurrence and distant metastases.³ Compared to other sarcomas, well-differentiated liposarcomas exhibit lower rate of local recurrence and significantly lower chance of distant metastasis.⁴ Traditional treatment includes surgical excision, often with concomitant organ resection. The literature is conflicting regarding the benefits of adding chemotherapy and radiation in a multimodality therapeutic regimen.² Despite aggressive treatment, patients experience a high rate of local recurrence and disease progression.⁴ We report a case of a 39cm retroperitoneal well-differentiated liposarcoma filling the entire right side of the abdomen, which was managed with surgery alone. The tumor was successfully excised with an organ-sparing approach which avoided radiation-related side effects and preserved renal function.

Case presentation

The patient is a 45-year-old man who presented with left scrotal swelling and pain. On physical examination he was found to have a large left hydrocele extending into the left inguinal canal. His comorbidities include hypertension, hyperlipidemia, obesity, and asthma; past surgical

history was significant for a right hydrocele repair the prior year, with normal scrotal ultrasound post procedure. CT of the pelvis confirmed the left hydrocele and identified a partially visualized large right retroperitoneal fat-containing mass, concerning for a retroperitoneal liposarcoma.

Subsequent MRI of the abdomen revealed a 26 × 19.5 × 16cm mass in the right retroperitoneum. Signal characteristics were consistent with a homogeneously fatty mass suspicious for low-grade liposarcoma (Fig. 1). Consensus recommendation by sarcoma and genitourinary multidisciplinary conferences was for preoperative radiation followed four to six weeks later by open midline right retroperitoneal mass resection, which is in accordance with current NCCN guidelines. Per NCCN guidelines, a biopsy was not recommended because diagnostic imaging was consistent with well-differentiated liposarcoma.⁵ The patient then met with the sarcoma multispecialty team, including radiation, medical and surgical oncologists. During radiation planning it became evident and was communicated to the patient that due to the location of his tumor, his right kidney would receive a full dose of radiation. As a result, his kidney was likely to be damaged and removed at the time of resection. Because of the patient's desire to preserve the functioning kidney, he elected to undergo surgical treatment without preoperative radiation. The patient underwent an open right retroperitoneal mass excision.

The resection approach consisted of a midline laparotomy. The tumor was mobilized off the surrounding organs, which included small

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<https://doi.org/10.1016/j.eucr.2020.101502>

Received 28 October 2020; Received in revised form 16 November 2020; Accepted 19 November 2020

Available online 24 November 2020

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Fig. 1. MRI of the patient's abdomen revealing a large $19.5 \times 16 \times 26$ cm right retroperitoneal lipoma, suspicious for a low-grade liposarcoma.

and large bowels, right kidney, right ureter and vena cava. In the right upper quadrant, the mass was resected widely with removal of all the perinephric fat, providing a wide resection in this area. Similarly, a wide resection was obtained posteriorly and laterally. Given the anatomic constraints, a wide resection was not achievable in all locations, such as where the tumor abutted the bowel and ureter. The patient experienced no complications and was discharged home on the second postoperative day. His renal function two days after surgery was consistent with preoperative levels (serum creatinine of 1.00mg/dL and eGFR of >60 mL/min/ 1.73 m²). Histology showed variable sized adipocytes with scattered enlarged and hyperchromatic atypical nuclei (Fig. 2), and focal cellular fibrous septa with atypical spindled cells. The tumor cells were positive for *MDM2* gene amplification via fluorescence in situ hybridization. The findings were those of a 39cm well-differentiated liposarcoma, which is a low-grade sarcoma. Margin assessment confirmed an R1 resection as he had grossly negative but microscopically positive margins. In the retroperitoneum, liposarcomas virtually always involve the margins microscopically. Eight-month follow-up showed no clinical

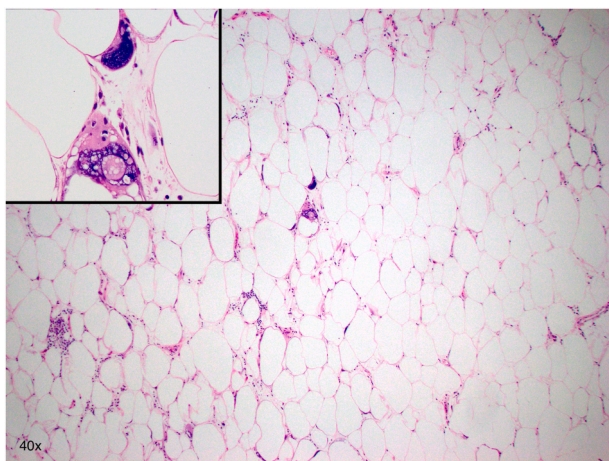


Fig. 2. Histology at low power magnification shows variable sized mature adipocytes with scattered enlarged and hyperchromatic nuclei (center). Top right insert shows high power magnification (200 \times) of the atypical hyperchromatic adipocyte nuclei.

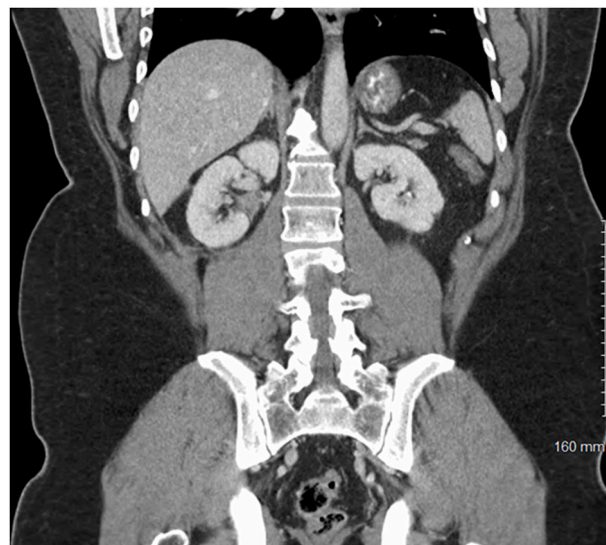


Fig. 3. Post-operative imaging of the patient's abdomen.

or radiographical evidence of recurrence (Fig. 3). Postoperative radiation was not recommended for the patient in accordance with NCCN guidelines.⁵ As the patient is at high risk for recurrence, which can occur years or even decades later, he will be closely monitored, both clinically and radiologically.

Discussion

Surgical management of retroperitoneal sarcoma is currently the gold standard. Data supporting preoperative radiotherapy is limited and has been justified by extrapolation from its well-established role in extremity sarcoma.² In this case, the patient's risk factors for developing chronic kidney disease and his concern to avoid renal disease or failure led to a treatment path which contradicted the multispecialty team's recommendation. Serendipitously, his desire to avoid radiation ended up in line with the results of a recently published multicenter, open-label, phase III randomized clinical trial (EORTC-62092:STRASS), which demonstrated similar abdominal recurrence-free survival at three years with surgery alone versus presurgical radiation followed by surgery.² The authors conclude that preoperative radiotherapy should not be considered standard of care for retroperitoneal sarcoma. In this series of 266 patients, the median tumor size was 19cm and more than 90% of patients underwent concomitant organ resection at surgery, including 80% performed with kidney resection.² Of note, 45% in the surgery alone group and 39% in the preoperative radiotherapy plus surgery group underwent systematic resection of the kidney in the absence of macroscopic involvement by cancer. A technique involving removal of the perinephric fat and Gerota's fascia without removal of the kidney may be feasible in many of these situations.

The present management seems particularly noteworthy as it was accomplished without nephrectomy or renal damage from radiation. Time will tell whether the surgical strategy to remove perinephric fat with preservation of the kidney itself will be sufficient to avoid abdominal recurrence in this patient. We suggest this as a surgical technique to be considered when a nephrectomy otherwise would have been performed. We would argue for a multidisciplinary surgical approach involving a urologic surgeon if this technique is not within the expertise of the primary oncologic surgeon.

Conclusion

Retroperitoneal sarcoma is an uncommon malignancy for which surgical excision without preoperative radiation is again the standard of

care, and should be strongly considered, especially in case of well-differentiated liposarcoma. Even in very large tumors, removing Gerota's fascia and perinephric fat with preservation of the kidney can result in macroscopically-clear margins, as demonstrated in this case. We suggest this as a surgical technique to be considered when a nephrectomy otherwise would have been performed, via a coordinated, multidisciplinary surgical approach if needed.

Author contributions

Joshua Kuperus: Writing- Original draft preparation, Reviewing and Editing. **Matthew Steensma:** Writing- Reviewing and Editing. **Vadim Khachaturov:** Visualization, Writing- Reviewing and Editing. **Brian Lane:** Conceptualization, Funding acquisition, Supervision, Writing- Reviewing and Editing.

Ethical approval

Spectrum Health Institutional Review Board does not require ethical approval for reporting an individual case report that only documents the observations of a patient receiving medical care because there is no intent to test a hypothesis via a systematic data analysis.

Declaration of competing interest

The authors have no conflicts of interest to declare.

Acknowledgment

The corresponding author would like to thank the Betz Family Endowment for Cancer Research for their support (RG0813-1036). Funding was provided in part by the Spectrum Health Foundation. The corresponding author would like to thank Sabrina Noyes for manuscript preparation and submission.

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