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Surgical challenges in the treatment of a giant renal cell carcinoma with atypical presentation: A case report



Rodolfo J. Oviedo (MD, FACS)^{a,b,*}, Jarrod C. Robertson (BS)^c, Kenneth Whithaus (MD)^d

^a Capital Regional Surgical Associates, 2626 Care Dr. Suite 206 Tallahassee, FL, 32308, USA

^b FSU College of Medicine, Clinical Assistant Professor of Surgery, 1115 W Call St. Tallahassee, FL, 32306, USA

^c FSU College of Medicine, 1115 W Call St. Tallahassee, FL, 32306, USA

^d Ketchum, Wood & Burgert Pathology Associates, 1899 Eider Ct. Tallahassee, FL, 32308, USA

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ABSTRACT

INTRODUCTION: For the management of localized renal cell carcinoma (RCC), surgical resection is the standard of care. Considerations are given to achieve good outcomes with conservative measures. When the tumor is exceedingly large the safest alternative is total nephrectomy.

PRESENTATION OF CASE: The patient is a 75 year old man with a 5 year history of increasing abdominal distension. There was no recent hematuria or any other genitourinary complaints. CT revealed a giant complex mass that occupied the majority of the abdomen likely arising from the retroperitoneum. Early in diagnosis, the mass was suspected to arise from the left kidney. The decision was made to proceed with surgery for both treatment and diagnosis. Resection of the tumor revealed a 28.0 × 25.0 × 15.0 cm encapsulated neoplasm. Histopathology determined this to be a papillary RCC. Resection of the mass resulted in en bloc partial nephrectomy immediately followed by a completion of the nephrectomy, lymphadenectomy, and abdominal wall repair. Postoperative course was excellent.

DISCUSSION: The aim of this report is to determine the surgical challenges posed by a tumor of this magnitude and the multidisciplinary approach necessary to treat it. In the often indolent course seen with RCC, surgeons are faced with the task of handling advanced disease, requiring more radical procedures for good outcomes.

CONCLUSION: The size of the tumor in this case presented several challenges in the operative setting. The sheer mass of the tumor gave no other choice than to perform exploratory laparotomy and complete nephrectomy upon resection.

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1. Introduction

RCC is the most common type of malignancy affecting the kidney [1]. Metastatic disease occurs in approximately 25%–30% of patients with renal cell carcinoma [2,3]. Because of the often indolent course of RCC, patients tend to present with advanced disease. Significant gains have been made recently in producing a number of new agents and approaches to the treatment of advanced RCC [4]. Nonetheless, this is a primarily surgical disease as the 5 year survival rate of patients without resection is approximately 10% [3]. This represents a 2.5 fold increase in overall and cancer-specific mortality when compared to surgical patients [5]. Surgery offers

several options for patients with RCC. For tumors without the presence of metastasis, conservative surgical resection is acceptable. For metastatic RCC there are two options. First, there exists cytoreductive nephrectomy for primary tumor resection with known metastatic burden. If cytoreductive nephrectomy is not an option or if additional debulking is desired, patients can undergo metastasectomy to remove distant foci of disease [5–9].

2. Presentation of case

This patient is a 75 year old man with a history of increasing abdominal distension (Fig. 1) that was recently accompanied by abdominal discomfort and constipation. He presented without any complaints of hematuria or other genitourinary symptoms. Physical exam showed a markedly protuberant abdomen with dullness to percussion and hyperactive bowel sounds. Computed tomography (CT) scan revealed a giant retroperitoneal mass possibly arising from the left kidney. On CT the mass measured 32.6 × 27.7 × 32.4 cm. Early in diagnosis, the differential also included a retroperitoneal sarcoma. However, the likelihood of a

Abbreviations: RCC (SPM314), renal cell carcinoma marker clone; CK7, cytokeratin 7.

* Corresponding author at: Capital Regional Surgical Associates, 2626 Care Dr. Suite 206 Tallahassee, FL, 32308, USA.

E-mail addresses: rodolfo.j.oviedo@gmail.com (R.J. Oviedo), Jcr09c@med.fsu.edu (J.C. Robertson), kwhit110@gmail.com (K. Whithaus).

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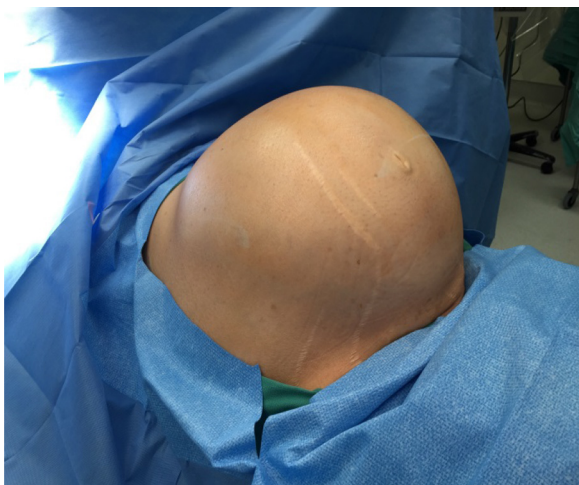


Fig. 1. Patient's abdomen prior to midline incision.



Fig. 2. Descending and sigmoid colon medialization for giant retroperitoneal tumor resection.



Fig. 3. Tumor just before complete resection.

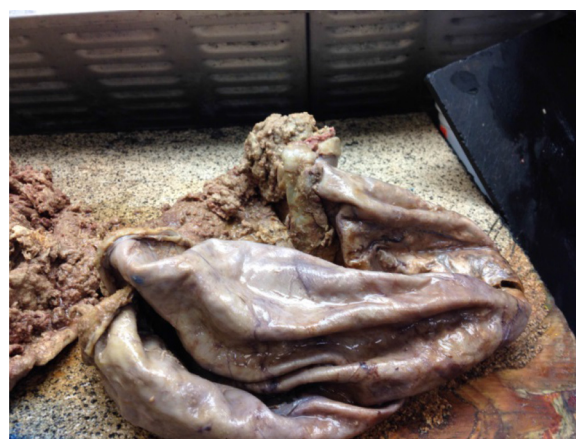


Fig. 4. Gross depiction of mass.

tumor arising from the left kidney was considered and guided early decision-making. Therefore, the decision was made to proceed with surgery for both treatment and diagnosis. Due to the size of this tumor, a multidisciplinary team was formed several weeks prior to surgery. Before laparotomy, bilateral ureteral catheters were placed by the urologist to help the surgeon identify and protect the ureters given the magnitude of this tumor. A plastic surgeon was on standby for reconstruction of the abdominal wall. A bowel prep was not given. He was kept on a liquid high protein diet three days prior to surgery. A midline incision was made and the peritoneal cavity was entered. The tumor was confirmed to be retroperitoneal. The descending colon was medialized from the rectosigmoid junction to the splenic flexure (Fig. 2). Care was taken to ensure the tumor capsule was preserved. A LigaSure electrothermal vessel sealing device (Valleylab, Boulder, CO) was used to divide the blood vessels supplying the tumor itself. The same approach was taken from the right side, mobilizing the hepatic flexure. The attachments to the tumor capsule were divided from the left side. At this point, we were made aware that as we were mobilizing the tumor, there was hypotension. Its weight caused significant compression of the inferior vena cava and decrease in preload. It took careful manipulation of the tumor to prevent persistent hypotension. Mobilization of the transverse and descending colon allowed for circumferential dissection from underlying structures and from the overlying mesentery. It was impossible to isolate the mass from the inferior pole of the left kidney, thus the kidney had to be sacrificed with an en bloc partial nephrectomy. The tumor was completely resected and handed off to pathology for frozen section (Fig. 3). Pathology

confirmed the presence of renal cell carcinoma with papillary features. The urologist was called back to perform a completion of the left nephrectomy and left periaortic lymphadenectomy. After that, the retroperitoneum was irrigated and all intraabdominal organs were examined and deemed unharmed including the right kidney and ureter. Because of the extent and duration of abdominal wall distension leading to skin redundancy and rectus diastasis, it was necessary for the patient to undergo reduction abdominoplasty in the hands of a plastic and reconstructive surgeon to achieve a good cosmetic and functional result with the closure. The patient was transported to recovery in good condition.

Histopathologic examination revealed a large grossly intact mass, 28.0 × 25.0 × 15.0 cm. The outer surface showed a partial peritoneal lining (Fig. 4). The tumor was opened to reveal abundant tan opaque fluid and necrotic material. Sectioning this area revealed a portion of possible ureter, 6.5 cm in length × 0.7 up to 2.5 cm in diameter. A thin rim of compressed renal parenchyma (representing the residual kidney) was confirmed microscopically.

Sections of the lesion revealed a neoplastic epithelial cell proliferation composed of pleomorphic cells in a predominantly papillary configuration (Fig. 5). There were scattered small swirled microcalcifications. While rare cells with clear cytoplasm were evident, these were a minority of the neoplasm. The grossly identified dilated ureter also appeared free of neoplastic growth. The tumor was positive for CK7 (OV-TL 12/30) and Renal Cell Carcinoma (SPM314) tumor markers. The para-aortic lymphadenectomy sections revealed multilobulated lymphoid tissue. There was an epithelial cystic lesion in one of the lymph nodes. Immunohisto-

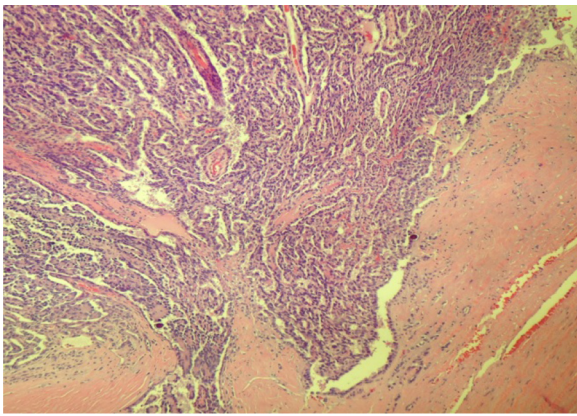


Fig. 5. Papillary renal cell carcinoma.

chemical evaluation showed a similar pattern to the malignant tumor. CK7 (OV-TL 12/30) and RCC (SPM314) tumor markers were positive.

The patient's postoperative course was uneventful except for a mild case of paralytic ileus which resolved after four days. His preoperative creatinine was 1.5 and it increased to 1.8 immediately postoperatively, but final creatinine on discharge was 1.3. His urine output was appropriate throughout the course of his recovery. The TNM classification was pT2bN1M0 (Stage III). Discussion with the patient and medical oncologist led to the conclusion that radical nephrectomy was the appropriate treatment based on National Comprehensive Cancer Network (NCCN) guidelines after positron emission tomography (PET) scan showed no other foci of disease. No adjuvant therapy was necessary at this time. Careful monitoring for relapse will be done with PET/CT every six months for two years and yearly thereafter.

3. Discussion

Surgical resection remains the gold standard treatment for RCC as a result of excellent oncologic outcomes [10]. The tumor resected in this case is a testament to the indolent course that can be seen with RCC. Fewer than 11% of RCC cases today present with the typical triad of hematuria, pain, and palpable mass [11,12]. Fortunately for this patient, despite the size of the tumor there was no evidence of metastatic disease determined by postoperative nuclear medicine whole body scan. Many advances have been made in the surgical approach to renal cell carcinoma, leading to more conservative treatments [7]. These options are null in cases such as this when tumor size is very large. In instances such as this when an open resection must be performed, anterior approaches are safest as they allow excellent visualization of the great vessels and renal hilum [10].

The size of this tumor presented unique challenges to the surgical team. To our knowledge, at $28 \times 25 \times 15$ cm and a total volume of $10,500$ cm³, this is the largest RCC in the world successfully resected and published in the English language. We performed an exhaustive literature review using the PubMed database and analysis of over 300 articles did not reveal a RCC tumor of this magnitude.

Despite advances in surgical technique, early detection and evaluation of RCC is still difficult, especially when symptoms like hematuria and other genito-urinary complaints are absent. Currently, up to 60% of all renal tumors are found incidentally, [7] with 20%–30% of RCC patients presenting with metastatic disease [8]. This factor, in addition to the indolent course of RCC, makes patients present at advanced stages of disease.

Although this was an atypical presentation of RCC given the lack of specific genitourinary symptoms, after discussion on Tumor

Board we committed to at least performing a palliative procedure to improve this patient's quality of life. Therefore, with a degree of uncertainty from a lack of specific preoperative diagnosis we performed the resection with the ability to adapt as the operation progressed. Proper planning was necessary in terms of securing a multidisciplinary approach in a 266-bed community based hospital. Some of these methods included insertion of preoperative ureteral catheters, the availability of a urologist to assist with completion nephrectomy after tumor resection by us, and the support from a plastic surgeon to reconstruct the abdominal wall depending on its condition at the end of the procedure. Lastly, perhaps the most challenging aspect of this case was the fact that the sigmoid and descending colon were being displaced by the tumor and had to be medialized with careful attention to protect their blood supply and preserve the mesentery, all while being cautious with the great vessels and ureters. Many general surgery exposure and mobilization maneuvers were employed with a successful outcome.

4. Conclusion

This case describes the treatment of the largest renal cell carcinoma ever reported in the literature. The often atypical nature of RCC poses a challenge to early diagnosis and presents surgeons with the monumental task of managing a significant disease that may be too advanced for minimally invasive approaches such as laparoscopy or robotic surgery.

Conflicts of interest

There are no conflicts of interest to report.

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Consent

Proper consent was obtained from the patient to publish writing and images pertaining to the case.

Author contribution

Rodolfo Oviedo M.D. FACS: Head of Study.
Jarrod Robertson MS3: main writer.
Kenneth Whithaus M.D.: writer.

Guarantor

Rodolfo Oviedo M.D. FACS.
Jarrod Robertson MS3.

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