International Journal of Medical and Pharmaceutical Case Reports
7(2): 1-5, 2016; Article no.IJMPCR.25900
ISSN: 2394-109X, NLM ID: 101648033

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Clinical Course of a Case with Sarcomatoid Renal Cell Carcinoma

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Authors' contributions

This work was carried out in collaboration between all authors. Authors OO and BA wrote the draft of the manuscript. Author AIH managed the literature searches. Authors AO and NCC designed the figures, managed literature searches and contributed to the correction of the draft. Author BN provided the case, the figures and supervised the work. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/IJMPCR/2016/25900
Editor(s):
(1) Rakesh Kumar Tiwari, Chapman University School of Pharmacy, Chapman University, Harry and Diane Rinker Health Sciences Campus, Irvine, CA, USA.
Reviewers:
(1) Cosimo Sperti, University of Padua, Padua, Italy.
(2) Georgios Tsoulfas, Aristotle University of Thessaloniki, Greece.
(3) Vital Hevia, Hospital Universitario Ramón y Cajal, Spain.
Complete Peer review History: http://sciencedomain.org/review-history/14323

Case Study

Received 25th March 2016
Accepted 19th April 2016
Published 25th April 2016

ABSTRACT

Introduction: Renal cell carcinoma (RCC) is the most frequently observed malign neoplasm of the kidney. Its sarcomatoid variant (SRCC) is rare but aggressive. Our aim is to present the clinical course of a 51 year-old patient with SRCC, in the light of the literature.
Methods: A patient applied to our clinic reporting weight loss, fatigue, flank pain, and abdominal mass. Magnetic resonance imaging revealed a hypointense mass that has a heterogenic T2-weighted outlook with a size of 95x110x147 millimeters. Patient underwent left radical nephrectomy, as well as colon resection and ileostomy due to invasion.
Results: Pathological diagnosis was reported as colon-invasive SRCC with a tumor sized 20x10x7.5 centimeters.
Conclusion: First resection has a key role because complete resections are the biggest chance of most patients for long-term survival.

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Keywords: Renal cell carcinoma; nephrectomy; metastasectomy; sarcoma.

1. INTRODUCTION

SRCC makes up 1-2% of all malign tumors of the kidney. It is more lethal than the carcinomas of prostate, bladder, and paratesticular regions [1]. Five-year survival rate is 29-36% and majority of patients die within the second postoperative year [2]. Similar to other large and fast growing RCCs, SRCC related symptoms include palpable masses, hematuria, stomachache, and flank pain. Findings, which suggest sarcoma rather than RCC, are origins in the capsule or perisinusoidal region, reaching big sizes without lymphadenopathy, lipid or bone structure indicating liposarcoma or osteosarcoma, and hypovascular qualities in angiography [3].

SRCC, a rare variant of renal tumors, is characterized by the microscopic presence of intercellular bridges or keratinization. Differentiating SRCC from renal sarcoma is difficult in spite of clinical and radiological findings, and even pathological evaluation in some patients. In general, most important prognostic factors for sarcomas are the condition of surgical border and degree of cancer.

Sarcomatoid differentiation is observed in 10-20% of renal cell carcinomas. Sarcamatoid components can be observed in all histological subtypes of RCC. Patients with sarcomatoid renal cell carcinoma are rare and are usually characterized by a rapidly progressing clinical course. In spite of the initial large excision and the recurrent mass excision on the postoperative 45th day, our case was deceased within the 7th post-operative month. Our aim is to discuss this rare case in the light of the literature.

2. PRESENTATION OF CASE

51-year-old male patient applied to our clinic reporting weight loss, fatigue, flank pain, and abdominal mass. Full abdomen magnetic resonance (MR) revealed a hypointense mass that has a heterogenic T2-weighted outlook with a size of 95x110x147 millimeters, partly cystic components, and heterogeneous intensity, which pushes the inferior-reaching kidney towards anterior by filling the left kidney region from the posterior. In the inferior neighborhood, although the border between them was not clear, there was a mass with heterogeneous intensity in the size of 113x126x124 millimeters, which had more cystic components. Loss of the normal signals of blood flow in renal vascular structures were observed. The spleen was pushed upwards. Colon, small intestine segments and aorta were pushed slightly to the right (see Fig. 1). Thorax CT and MR imaging did not display any metastasis. Patient underwent left radical nephrectomy with chevron incision, as well as colon resection and ileostomy due to invasion. There was no palpable lymphe node so lymphadenectomy was not performed.

Pathology specimen revealed Pansitokeratin, Vimentin, Masson-Trichrome, S-100, CD-10, CD-31, Desmin, Myo-D1, and positive reaction in the tumor cells. Pathological diagnosis was reported as colon-invasive SRCC (pathological stage 4 according to the TNM classification) with a tumor sized 20x10x7.5 centimeters, Fuhrmann nuclear grade 4. In addition to SRCC, tumor tissue displayed conventional (clear cell) RCC, fibrosarcoma, leiomyosarcoma, and anjiosarcoma morphology, and partially included hyalinized and necrotic calcification and bleeding areas. Surgical margin was reported as negative and vascular invasion was not determined in the pathology specimen. Tumor invasion to colon's serosal surface was observed. Invasion consisted of tumor's sarcomatoid component.

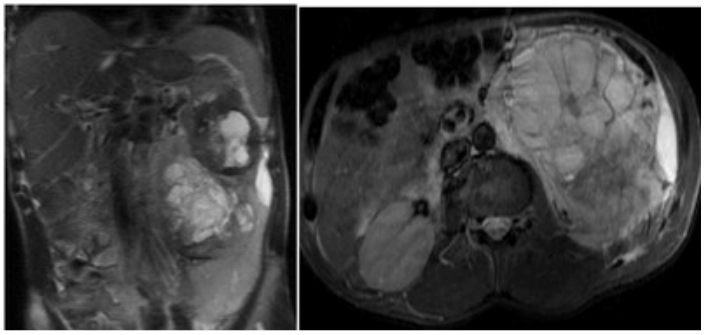

Fig. 1. MRI images of the renal mass

Patient was discharged on the sixth postoperative day with ileostomy, and was referred to medical oncology. 45 days after the first operation in the ultrasound imaging performed before planned chemotherapy, revealed a recurrent mass in the abdomen. In the contrasted computed tomography of the abdomen, there was a 14x7.6-centimeter mass image, filling the left kidney region and left infracolic compartment. The mass was regularly contoured, had parts with enhanced contrast and was hard to clearly differentiate from the anses of the ileum (see Fig. 2). The patient was consulted to uro-oncology council (urologist, medical and radiological oncologist, radiologist) and the council was decided to exploration. The previous incision track was used to reach the recurrence mass. The mass was invasive to ileum at 3 region and it was excised with adjacent parts of ileum. Pathology results were consistent with SRCC (Fuhrmann nuclear grade 4) infiltration.

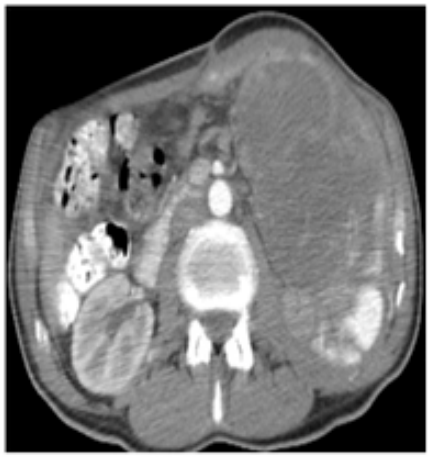

Fig. 2. CT image of the recurrent mass

Patient was discharged on the 9th postoperative day. After the second resection, the patient began medical oncology treatment with doxorubicin and gemcitabine, which are both chemotherapeutic agents.

Contrast MRI of the full abdomen a month after the second resection revealed lobular-contoured solid mass lesion sized approximately 93x41 millimeters (see Fig. 3).

Due to severe disturbance in the patient's general health status, a third resection was not performed. The patient was deceased 6.5 months after the initial diagnosis.

3. DISCUSSION

Classification of World Health Organization (WHO) defines SRCC as a histological subtype of RCC containing high grade malign fusiform cells. SRCC, with its mesenchymal (sarcomatosis) and epithelial (carcinomatosis) agents, is a biphasic legion [4]. Because of having an average six-month survival rate, it is almost accepted as a deadly cancer. Our patient's overall survival was 6.5 months in accordance with the literature. Congiano and colleagues have conducted a case series of 31 patients, and reported first and second year survival rates as 48% and 36% respectively, when surgical resection is combined with immunotherapy [5].

High-grade sarcomas usually metastasize and majority of patients die within months due to progression of the disease. Low-grade sarcomas, on the other hand, tend to have a slower progression. However, due to local recurrences, recurrent resections are required to extend survival and decrease morbidity [6]. Imaging of our patient did not reveal any solid organ (lung, bone, liver, brain etc.) metastasis, and he survived for 6.5 months.

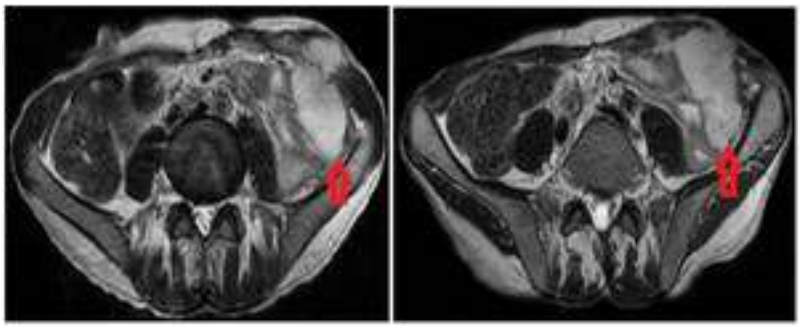

Fig. 3. MRI images of the recurrent mass

Imaging qualities unique to SRCC are especially limited. MRI can be efficient for detecting sarcomatoid differentiation. SRCC displays an irregular or infiltrative image on MRI, and there is heterogeneous signal intensity in T2 imaging. Clinical symptoms of SRCC are not different than all other RCCs. Symptoms associated with SRCC can be due to local tumor growth (hematuria, flank pain, abdominal mass, perirenal hematoma), hemorrhage, paraneoplastic syndromes (hypercalcemia, hypertension, polycythemia) or metastatic disease (persistent cough, bone pain, cervical lymphadenopathy, constitutional symptoms, weight loss, fever, malaise. Capsulated tumor, lack of invasion to surrounding tissues, and low mitotic activity are accepted as good prognostic factors in some cases. However, even these good prognostic factors failed to reverse the aggressive clinical progress following the nephrectomy.

In general, most important prognostic factors for sarcomas are condition of the surgical border and degree of cancer. Recent studies have also reported percentage of sarcomatoid differentiation plays an important role in prognosis, with sarcomatoid differentiation above 25% having a negative effect. Each time the sarcomatoid differentiation is up by 10%, SRCC-related deaths also increase by 6%, and 30% was detected to be a significant cut-off score [7].

Systemic therapy for patients with SRCC has routinely been ineffective. Sella et al. found that patients who received a doxorubicin-based regimen were the only long-term survivors. Responses have also been reported using the MAID regimen (sodium mercaptoethanesulfonate, doxorubicin, ifosfamide, and dacarbazine), gemcitabine, docetaxel and carboplatin. In contrast, no responses were observed in patients receiving doxorubicin and ifosfamide [8,9]. Some studies report an extension by 10.9 months in the median survival rate when the patients are treated with sorafenib after the gemcitabine and doxorubicin [10]. There are also reports of 15.7-months average survival rate with sunitinib, and one study using VEGF tyrosine kinase inhibitors has reported an average of 11.8 months [11].

Even though our patient had no metastasis at the time of diagnosis, and had negative surgical borders, recurrence had occurred on the 45th postoperative day and this giant mass was resected. Another mass was detected prior to chemotherapy, but resection was avoided due to severe disturbances in the patient's general health status. Patient was exitus 6.5 months after the surgery. Patients with a potential diagnosis of SRCC should undergo radical nephrectomy in the early phase, and chemotherapy during the postoperative phase. However, in spite of the available aggressive and radical treatments, results are not promising.

4. CONCLUSION

First resection has a key role because complete resections are the biggest chance of most patients for long-term survival. For renal sarcomas, this resection includes radical nephrectomy and en-block removal of surrounding tissues.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
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