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CASE REPORT

Diagnosis and Management of Renal Sarcoma

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ABSTRACT

This is a case study related to renal sarcoma. Male patient of 66 years was studied who presented complaining of left flank pain. The images showed suspicious cystic mass, he underwent Robotic radical nephrectomy. The histopathology came as renal sarcoma. The concept of renal sarcoma has been presented in details, literature has been reviewed related to diagnosis, prognosis and case studies related to the subject done by different researchers in the field. The case has been presented in detail and discussed thereafter.

Introduction

Cancer of the soft tissues of the kidneys, the connective tissue or the fat around the kidneys is called renal sarcoma and is very uncommon. Renal sarcomas account for a very small percentage (1%) of all kidney malignancies (Argani, Pedram, et al. 2020). Renal sarcoma, like many other cancers, may develop when there are alterations to the DNA that makes up your genes. Cancer-causing DNA mutations often disrupt both oncogenes (which regulate cell division and survival) and tumor suppressor genes (which help to keep cell division in check). Unchecked growth of abnormal cells in either of the kidneys is renal (kidney) cancer. Renal sarcoma is an uncommon kind of kidney cancer that develops in the kidney's blood vessels or connective tissue. Only around 2% of primary kidney malignancies are renal sarcomas (Argani, Pedram, et al. 2019). The poor prognosis of fibrosarcomas is mostly attributable to their late detection, as well as the occurrence of locally progressed involvement into the renal vein or metastatic disease at the time of first examination. Fewer than 20% of patients survive after five years (Stahl, Christopher C., et al. 2021). It's also possible to develop less common types of sarcoma, such as leiomyosarcoma, rhabdomyosarcoma, osteogenic sarcoma, or liposarcoma. Eighty percent of patients will have a palpable mass in their abdomen, and forty percent will also be experiencing abdominal discomfort (Blanc, Thomas, et al. n.d.). Their main common presenting symptoms are hematuria, anemia, hypertension, and abrupt severe abdominal discomfort. An abdominal ultrasound is an impotent diagnostic tool for the diagnosis to start with and specially to differentiate the cystic from solid mass. CT scan should also be performed to for more tumor characteristic also to see the extension of the tumor and to rule out any metastasis. Histopathology is the gold standard for making the diagnosis. Together, surgical, radiation, and chemotherapy have resulted in impressively high percentages of cure. To the extent possible, the whole tumor should be removed surgically.

Literature review

Renal sarcomas don't seem to have a predilection for extending into the venous system or the inferior vena cava, however Shirkhoda and Lewis (1987) observed that sarcomas originating in the renal parenchyma and SRCC cannot be clearly differentiated from renal cell carcinoma. Adolescent renal sarcoma accounted for 0.8% of kidney cancer cases and carries a poor prognosis, according to research by Wang, Xianding, et al. (2011). The prognosis for patients improves with

early detection and surgical resection (Iorga, Lucian, et al. 2018)

Studying a 39-year-old male, Yang, Jun Ho, et al. (2020) showed that preoperative detection of major pulmonary tumor embolism linked with renal neoplasms is unusual. Emboli from a tumor in the lung are usually discovered after surgery to remove a kidney tumor. The incidence of primary renal sarcoma is low, and the incidence of primary renal sarcoma accompanied by pulmonary tumor embolism is much lower (Manikandan, Ramanitharan, et al. 2019). They present a case of renal sarcoma with a tumor embolus in the pulmonary artery. So, they reasoned, individuals with renal sarcoma who also have a pulmonary tumor embolism would benefit from chemotherapy followed by concurrent cytoreductive nephrectomy and pulmonary embolectomy.

Among adult patients with renal sarcoma, Daniel M. Moreira et al. (2015) evaluated the relationship between clinicopathologic features, treatment mode and survival. Around 8 percent of patients with metastatic renal sarcoma survive 5 years. The 5-year survival rate for people with non-metastatic renal sarcoma is 46%. Survival from renal sarcoma is correlated with patient age, race, tumor size, and tumor grade. Metastatic renal sarcoma has a dismal outlook, although localized tumors have a chance of being successfully treated and resulting in long-term survival (Michael, Aleisha E., et al. 2019).

Researchers Roberto Iacovelli et al. (2012) established a correlation between the clinical and pathological aspects of primary renal synovial sarcoma. It was observed that the likelihood of recurrence for individuals with non-metastatic illness at diagnosis was 36%, and that median overall and disease-free survival had been recorded. Primitive kidney tumors exhibited a wide variety of histological sub-types that correlated with tumor extent at diagnosis, immunohistochemical staining patterns, and genetic alterations, much as has been observed in other synovial sarcomas. Although this investigation was conducted retrospectively, the authors observed that the various forms of renal sarcomas are distinguished by distinct patterns of immunohistochemistry staining and translocations. Even while one in three individuals with non-metastatic illness had disease recurrence, the prognosis was much worse for those identified at the metastatic stage. To further characterize prognosis and treatment solutions for this uncommon condition, we need collaborative efforts and the publishing of cases with appropriate follow-up.

According to research by Bakhshi, Girish D., et al. (2012), synovial sarcomas are rare, deep-seated cancers that develop around the major joints of

young adults and adolescents. The prognosis for primary synovial sarcoma of the kidney is dismal because of its rarity and difficulty to treat (Argani, Pedram, et al. 2018). Immunohistochemistry is the sole method for making a diagnosis. If you have a sarcomatoid or spindle cell tumor, this might be a possible differential diagnosis. This case study focuses on a 33-year-old woman who had a radical nephrectomy performed on her left kidney due to a renal tumor. Histopathology and genetic studies identified it to be primary renal synovial sarcoma. Radiation treatment was administered, and the patient's progress has been monitored for two years with no complications.

The purpose of the research by Verheijen, Remy B., et al. (2017) was to investigate the pharmacokinetic and exposure-survival associations of pazopanib in a clinically relevant patient population. Inclusion criteria comprised individuals with renal cell carcinoma or soft tissue sarcoma for whom at minimum one pazopanib plasma concentration was obtainable. The results of this investigation support the hypothesis that a pazopanib $C_{min} > 20$ mg/L is associated with improved progression-free survival in kidney cancer, and they hint to a similar tendency in sarcoma patients. Pazopanib C_{min} monitoring may assist doctors determine whether a greater dosage is needed for a certain patient (Brillantino, C., et al. 2019).

Researchers Nazemi and Daneshmand (2020) observed that bladder and kidney sarcomas were the most prevalent types of genitourinary sarcomas. Sarcomas of the bladder and kidney had lower survival than paratestis and scrotum. Among bladder cancers, leiomyosarcoma had the longest

median survival time. The median survival period for kidney tumours was longer for liposarcomas. Kidney and paratesticular leiomyosarcomas are the most prevalent sites of presentation. Bladder, kidney, paratestis, and scrotum tumors are the most prevalent types, with kidney sarcomas having particularly poor prognoses. Treatment methods should be modified by type of sarcoma and initial tumor site within the GU tract since survival of comparable histologic types differed by primary tumor location (El Chediak, Alissar, et al. 2018).

The researchers (Zhu, Yongkun, et al., 2022) set out to create a predictive nomogram model that may predict OS and cancer-specific survival (CSS) in adult patients with RS. The study's authors developed and tested a predictive nomogram to help urologists determine a patient's likely outcome from renal sarcoma in adults.

Case

In our research we present the case of a 66 years old male patient who was presented with a history of diabetes mellitus, hypertension and coronary heart disease status post PCI.

He presented to the regional hospital few months before his presentation to our hospital, complaining of recurrent attack of left flank pain. There was no associated symptoms were reported. Initial investigation was done including ultrasound, which showed left lower pole hypoechoic complicated cystic mass measuring around 6.3 x 6.2 cm.

The ultrasound was done for the patient to analyse the state of the renal sarcoma and its growth in the patient. The figure 1 shows the ultrasound picture for the patient.

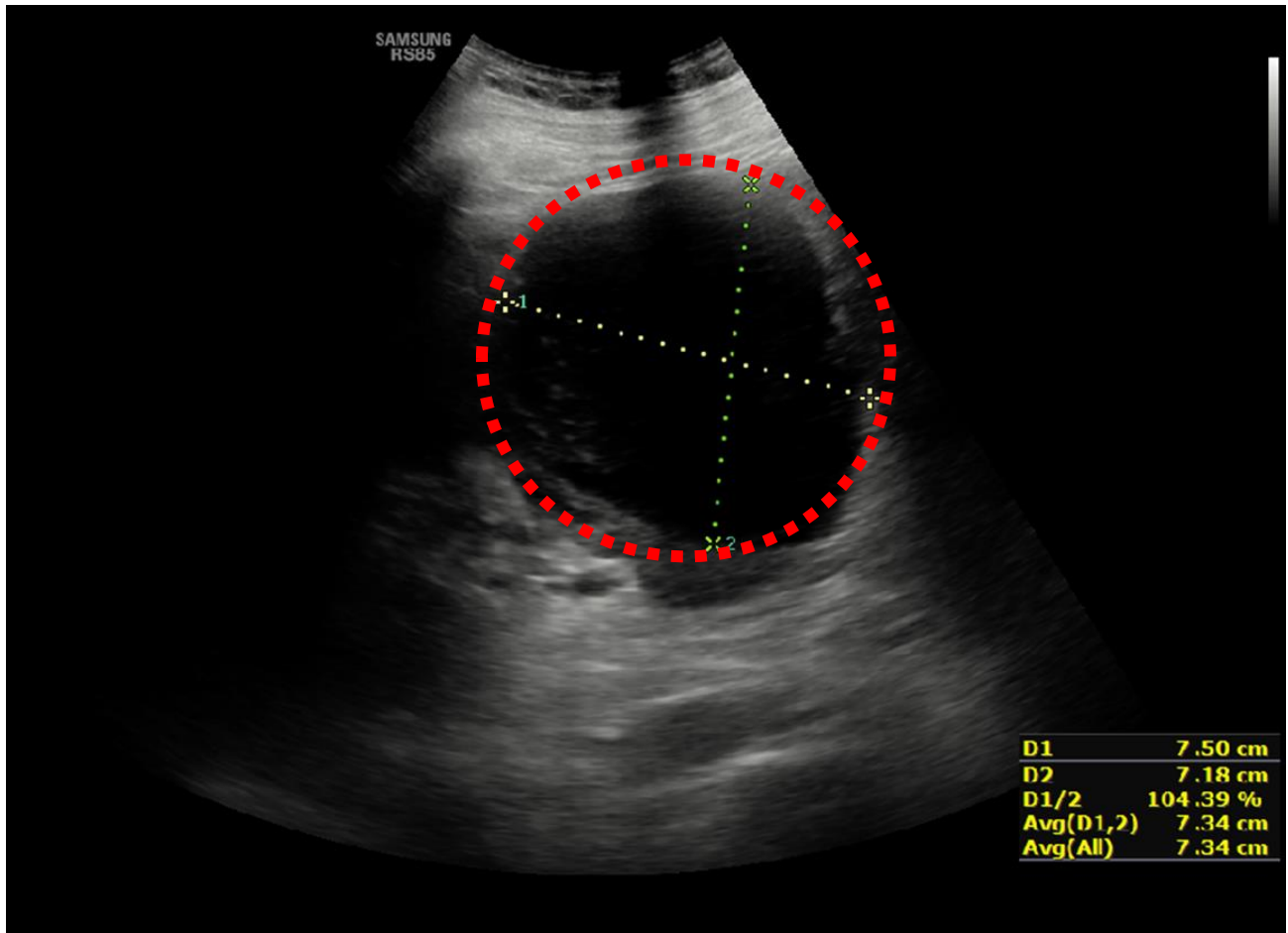


Figure 1. Showing ultrasound of the patient that revealed that he was having 6.3 x 6.2 cm cystic mass

CT urogram was done which showed left renal cystic mass measuring around 6 x 6 cm defined as a complicated cyst going with Bosniak grade 3 with

highly suspicion for malignancy. MRI was done, which showed left renal mass measuring around 7 x 8 cm with altered signal intensity .



Figure 2. CT Urogram coronal imaging.

The patient then was referred to our facility for further management. Upon his visit to us the CT scan was repeated and it showed a left complicated cystic renal mass measuring around 9 x 8.4 x 7 cm.

Computed tomography (CT) scans confirmed the presence of a big tumor inside the left kidney, which was consistent with renal cancer.

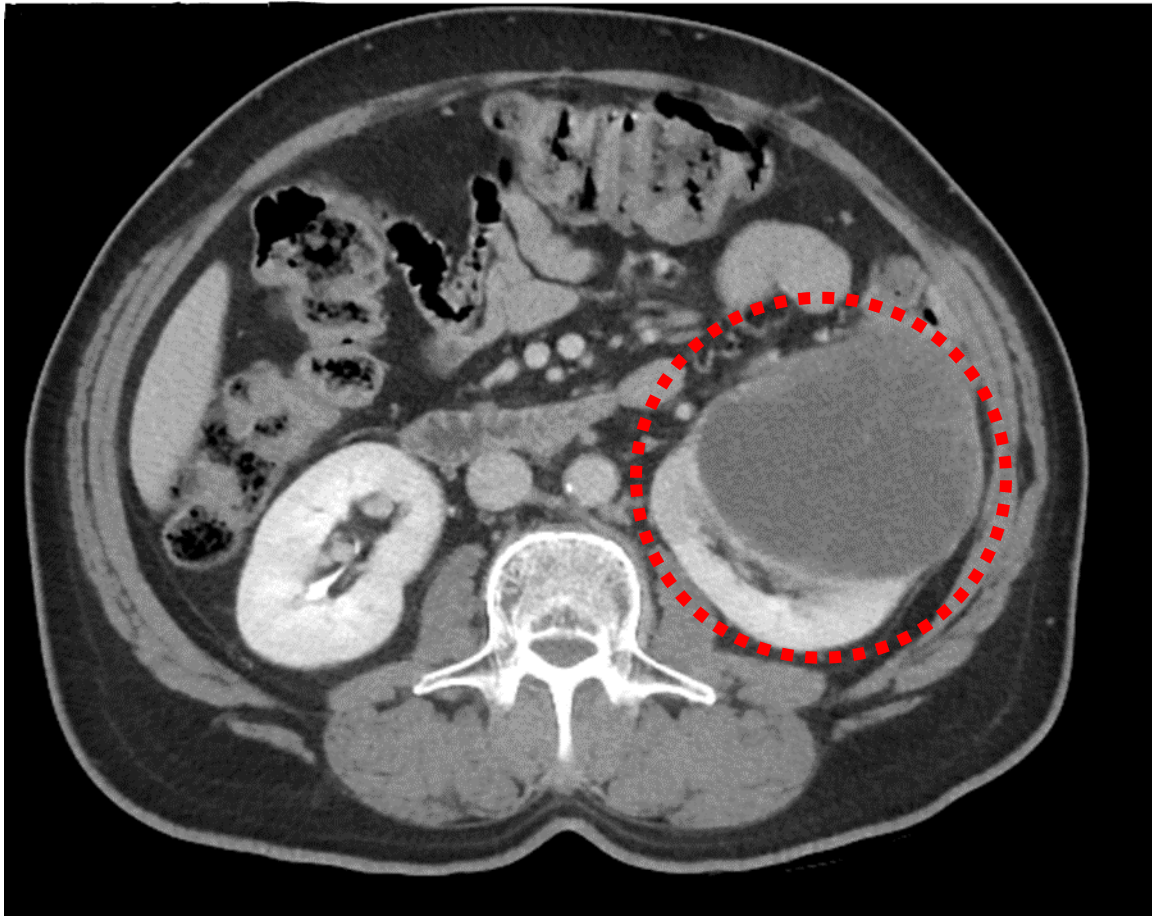


Figure 3. Showing CT scan with contrast, axial section

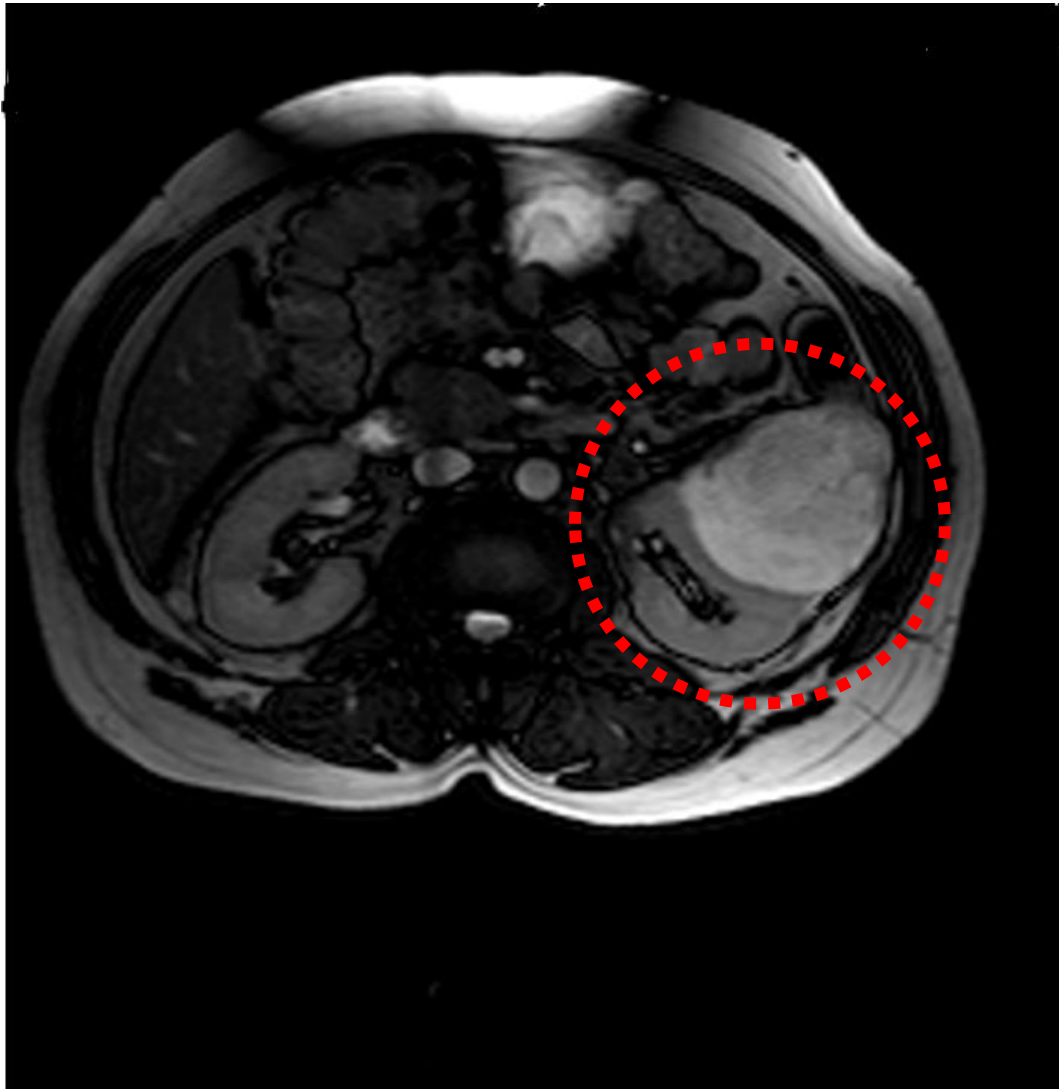


Figure 4. MRI of abdomen , axial imaging

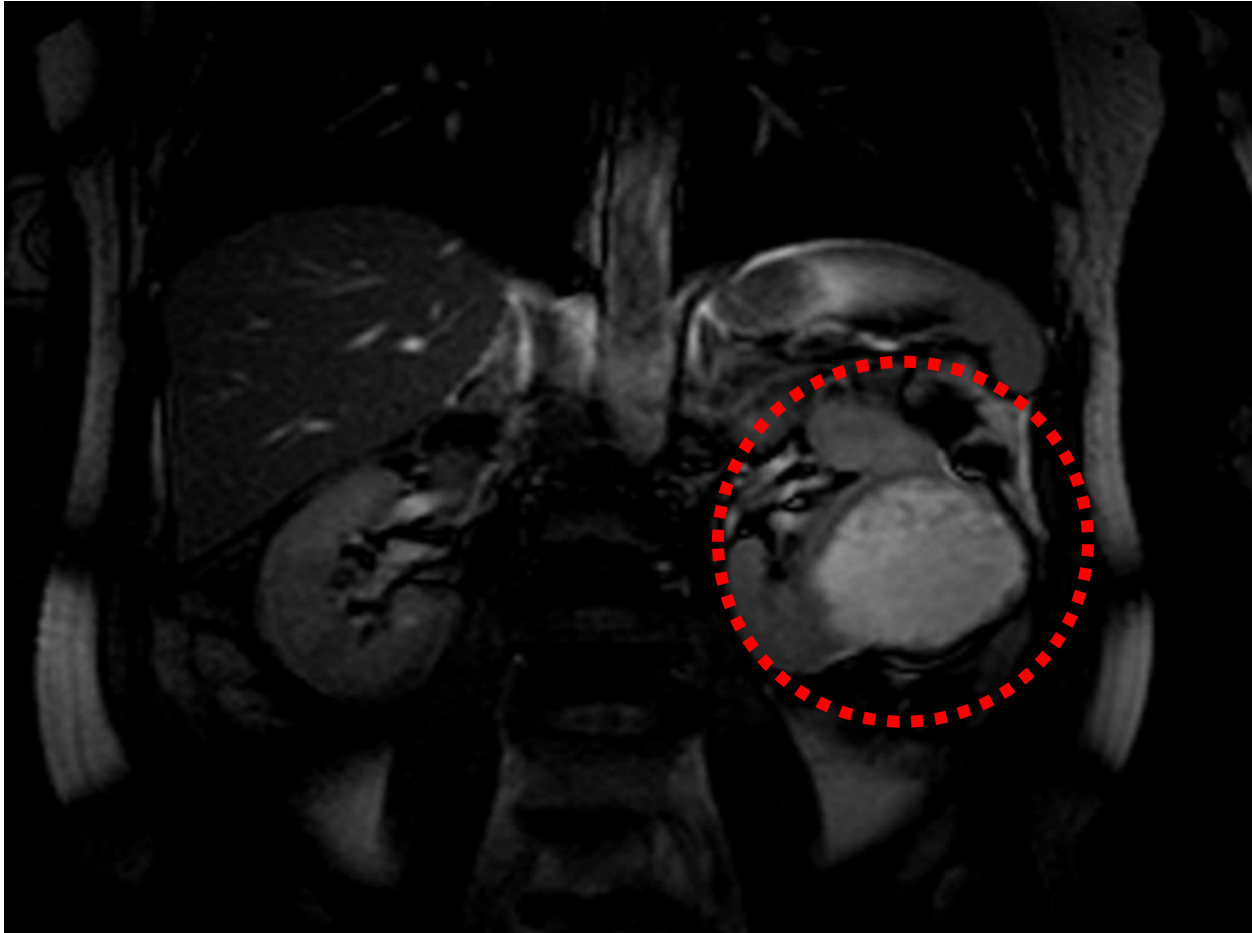


Figure 5. MRI of the abdomen, coronal section

The patient underwent robotic left radical nephrectomy which was done uneventful. The patient was discharged from the hospital on day two in a good and stable condition.

Histopathology of the cystic renal mass came as high grade pleomorphic sarcoma with surgical free margin and it was revealed that it was renal sarcoma. The patient is currently following with oncology surface in his regional hospital.

The patient was following back to the local hospital in his area and he was following up through imaging scans through CT and MRI. He was managed by the local hospital after the surgery and any requirement for the adjutant surgery.

The patient was not followed up at this hospital after the completion of his surgery and he was referred back to the local hospital in his area.

Discussion

In this case surgical intervention to remove the cyst was the best possible treatment. Size, differentiation, and tumor grade are all factors in determining prognosis in renal sarcoma. In order to get the best possible results, first surgical resection

with negative margins is essential. The best course of action for treating this uncommon and aggressive tumor is radical surgical resection with broad negative margins, followed by clinical follow-up to check for tumor recurrence. The size of the tumor was relatively large but it was found that it was not extending beyond the kidney. The patient was then referred to the oncologist for further management if need for chemo radiation. Small, low-grade sarcomas are often curable with surgical excision alone. Surgery and radiation treatment are typically used to treat those that are of high grade and greater than 2 inches (5 cm). The tumour may be shrunk with radiation treatment or chemotherapy prior to surgery. Here in this case the size was significantly large so patient was referred for the chemo radiations for further management. In the case of low-grade, early-stage sarcomas, surgery is often the only course of therapy necessary. However, further treatments, such as chemotherapy, radiation therapy, targeted therapy, or immunotherapy, may be necessary for more advanced sarcomas. We also consulted with a nutritionist to help him manage his diet to recover early. The nutritionist recommended him Zinc rich

diet, which is useful in promoting wound healing, and it can be found in chicken, beef, shellfish, and beans. Finally, we also recommended that he should be getting enough vitamin C by eating foods like broccoli and strawberries, which are naturally rich in the vitamin. The two together should hasten the process of healing wounds. The patient was recovering fast and he was stable as per the last update.

Conclusion

Primary renal sarcoma, in conclusion, is an aggressive uncommon illness that has characteristics

with other forms of renal cell carcinomas. Rare and deadly, renal sarcoma lacks of standard treatment protocols. Because of the aggressive nature of this illness and its propensity to return, we think that wide surgical resection with negative margins may be the best way to manage this condition with possible adjuvant chemotherapy or radiation therapy. Monitoring for tumor recurrence via follow-up is also crucial.

References

1. Argani, P., Kao, Y. C., Zhang, L., Sung, Y. S., Alaggio, R., Swanson, D., ... & Antonescu, C. R. (2019). BCOR overexpression in renal malignant solitary fibrous tumors: a close mimic of clear cell sarcoma of kidney. *The American journal of surgical pathology*, 43(6), 773.
2. Argani, P., Reuter, V. E., Kapur, P., Brown, J. E., Sung, Y. S., Zhang, L., ... & Antonescu, C. R. (2018). Novel MEIS1-NCOA2 gene fusions define a distinct primitive spindle cell sarcoma of the kidney. *The American journal of surgical pathology*, 42(11), 1562.
3. Argani, P., Zhang, L., Sung, Y. S., Bacchi, C., Swanson, D., Dickson, B. C., & Antonescu, C. R. (2020).
4. Bauer, F., Schmidt, T., Eisfeld, H., Dubois, C., Kastrati, K., Hochhaus, A., & Huebner, J. (2018). Information needs and usage of complementary and alternative medicine in members of a German self-help group for gastrointestinal stroma tumours, sarcoma, and renal cancer. *Complementary Therapies in Medicine*, 41, 105-110.
5. Blanc, T., Pio, L., Clermidi, P., Muller, C., Orbach, D., Minard-Colin, V., ... & Sarnacki, S. (2019). Robotic-assisted laparoscopic management of renal tumors in children: Preliminary results. *Pediatric Blood & Cancer*, 66, e27867.
6. Brillantino, C., Rossi, E., Minelli, R., Irace, D., Castelli, L., Zeccolini, R., ... & TUFANO, A. (2019). A rare case of renal tumor in children: clear cell sarcoma. *Il Giornale di Chirurgia-Journal of the Italian Surgical Association*, 40(3), 217-224.
7. Novel SS18-NEDD4 gene fusion in a primary renal synovial sarcoma. *Genes, Chromosomes and Cancer*, 59(3), 203-208.
8. Bakhshi, G. D., Khan, A. S., Shaikh, A. S., Khan, M. A. A., Khan, M. A. A., & Jamadar, N. M. (2012). Primary renal synovial sarcoma. *Clinics and Practice*, 2(2), e44.
9. El Chediak, A., Mukherji, D., Temraz, S., Nassif, S., Sinno, S., Mahfouz, R., & Shamseddine, A. (2018). Primary synovial sarcoma of the kidney: a case report of complete pathological response at a Lebanese tertiary care center. *BMC urology*, 18(1), 1-9.
10. Iacovelli, R., Altavilla, A., Ciardi, A., Urbano, F., Manai, C., Gentile, V., & Cortesi, E. (2012). Clinical and pathological features of primary renal synovial sarcoma: analysis of 64 cases from 11 years of medical literature. *BJU international*, 110(10), 1449-1454.
11. Iorga, L., Anghel, R., Marcu, D., Spinu, D., Pantea Stoian, A., Diaconu, C., ... & Bratu, O. G. (2018). Renal sarcoma—a rare parenchymal tumor with a very poor prognosis. *Arch Balk Med Union*, 53(3), 434-438.
12. Manikandan, R., Mehra, K., Dorairajan, L. N., Ganesh, R. N., Sreenivasan, S. K., & Kumar, R. (2019). Case Report: Partial nephrectomy in primary renal sarcoma presenting as Wunderlich syndrome; a rare tumour with rare presentation managed atypically. *F1000Research*, 8.
13. Michael, A. E., Grimes, J. A., Volstad, N. J., Osekavage, K. E., & Koenig, A. (2019). Inappropriate Secondary Erythrocytosis in a Dog With Renal Sarcoma. *Topics in companion animal medicine*, 36, 9-11.
14. Moreira, D. M., Gershman, B., Thompson, R. H., Okuno, S. H., Robinson, S. I., Leibovich, B. C., & Boorjian, S. A. (2015, December). Clinicopathologic characteristics and survival for adult renal sarcoma: A population-based study. In *Urologic Oncology: Seminars and Original Investigations* (Vol. 33, No. 12, pp. 505-e15). Elsevier.
15. Nazemi, A., & Daneshmand, S. (2020, May). Adult genitourinary sarcoma: a population-based analysis of clinical characteristics and survival. In *Urologic Oncology: Seminars and Original Investigations* (Vol. 38, No. 5, pp. 334-343). Elsevier.
16. Shirkhoda, A., & Lewis, E. (1987). Renal sarcoma and sarcomatoid renal cell carcinoma: CT and angiographic features. *Radiology*, 162(2), 353-357.
17. Stahl, C. C., Schwartz, P. B., Ethun, C. G., Marka, N., Krasnick, B. A., Tran, T. B., ... & Abbott, D. E. (2021). Renal function after retroperitoneal sarcoma resection with nephrectomy: a matched analysis of the United States sarcoma collaborative database. *Annals of Surgical Oncology*, 28(3), 1690-1696.
18. Verheijen, R. B., Swart, L. E., Beijnen, J. H., Schellens, J. H., Huitema, A. D., & Steeghs, N. (2017). Exposure-survival analyses of pazopanib in renal cell carcinoma and soft tissue sarcoma patients: opportunities for dose optimization. *Cancer Chemotherapy and Pharmacology*, 80(6), 1171-1178.
19. Wang, X., Xu, R., Yan, L., Zhuang, J., Wei, B., Kang, D., ... & Lu, Y. (2011). Adult renal sarcoma: clinical features and survival in a series of patients treated at a high-volume institution. *Urology*, 77(4), 836-841.
20. Yang, J. H., Song, D. H., Lee, C., Kang, D. H., Jung, J. J., Kim, S. H., ... & Moon, S. H. (2020). Malignant pulmonary embolism associated with renal sarcoma: A case report. *Medicine*, 99(19).

21. Zhu, Y., Mao, W., Zhang, G., Sun, S., Tao, S., Jiang, T., ... & Chen, M. (2022). Development and validation of a prognostic nomogram for adult patients with renal sarcoma: A retrospective study based on the SEER database.