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Case report

# A rare case of bilateral synchronous renal cancer, characterized by distinct histologies; leiomyosarcoma and clear cell renal cancer

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ABSTRACT

*Introduction:* Renal cancer is a significant global cause of death and clear cell being the most common subtype. Bilateral synchronous renal cancers with different histologies are extremely rare and less reported. Managing bilateral renal cancer is challenging.

*Clinical presentation:* A 51-year-old woman with a history of hypertension and hypothyroidism presented with right loin pain, leading to the discovery of a 7 cm right renal mass and a 2.3 × 2.6 cm mass on the upper pole of the left kidney, associated with reactive right para-aortic lymph nodes. The right kidney mass was identified as leiomyosarcoma with liver metastasis while the left kidney mass was diagnosed as clear cell renal carcinoma. Patient was managed with right side nephrectomy, left side microwave ablation and follow-up chemotherapy.

*Discussion:* Primary leiomyosarcoma of kidney is extremely rare and aggressive, leading to poor outcome. Synchronous bilateral renal cancer pose surgical challenges due to potential loss of renal function. In this case of differing histologies in synchronous bilateral renal masses, a nephron-sparing approach was taken, but despite aggressive treatment, the patient developed metastases in the liver and peritoneum.

*Conclusion:* Bilateral synchronous renal cancer, particularly with differing histologies in each kidney, are exceptionally uncommon, making their management challenging. Despite the limited guidance available due to their rarity, addressing these cases aggressively and promptly is crucial, as their prognosis is generally unfavorable, necessitating further research to advance management strategies.

## 1. Introduction

Renal cancer is responsible for approximately 180,000 deaths worldwide, making it a relatively common form of cancer. Each year, more than 430,000 new cases are diagnosed [1]. Significant advancements in the histopathological and molecular characterization of renal cell carcinoma (RCC) have led to its classification. Among the major subtypes, clear cell renal cell carcinoma (ccRCC), papillary RCC (pRCC), and chromophobe RCC (chRCC) stand out, while the remaining subtypes are exceptionally rare. In cases where tumors do not align with these classifications, they are categorized as unclassified RCC (uRCC). Clear cell RCC constitutes the most frequently identified histological type in RCC [2]. Primary renal sarcomas make up 0.8 % to 2.7 % of all renal tumors in adults, and among renal sarcomas, leiomyosarcoma is the predominant histological subtype, accounting for 50 % to 60 % of cases. Renal sarcomas can originate from various kidney components, including the smooth muscle fibers of the renal parenchyma, renal capsule, renal pelvis, and renal vessels [3]. The synchronous diagnosis of bilateral renal cancers (RCCs) occurs in 1 % to 5 % of cases [4]. Managing bilateral renal masses presents a surgical challenge, with the goal being the preservation of renal mass without resorting to dialysis [5]. This report was drafted in line with the SCARE 2020 criteria [6].

## 2. Case presentation

A 51-year-old female with a medical history of hypertension and hypothyroidism presented with persistent right loin pain lasting for 2 months. She did not report any lower urinary tract symptoms (LUTS),

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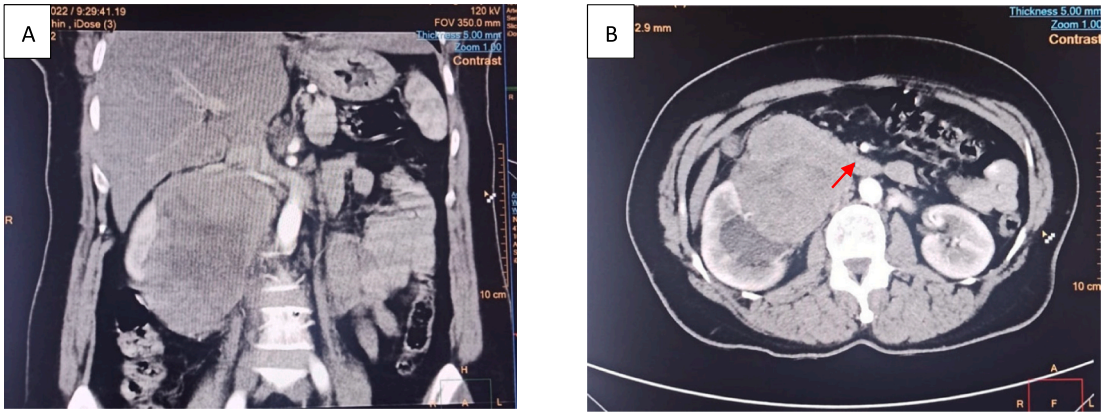


**Fig. 1.** Contrast enhanced computed tomography (CECT) of chest, abdomen and pelvis showing right side renal mass. (A) CECT image showing a heterogeneously enhancing mass with central necrosis involving the entire kidney, including the renal pelvis. (B) CECT image showing the tumor extension into the right renal vein (indicated by the arrow).

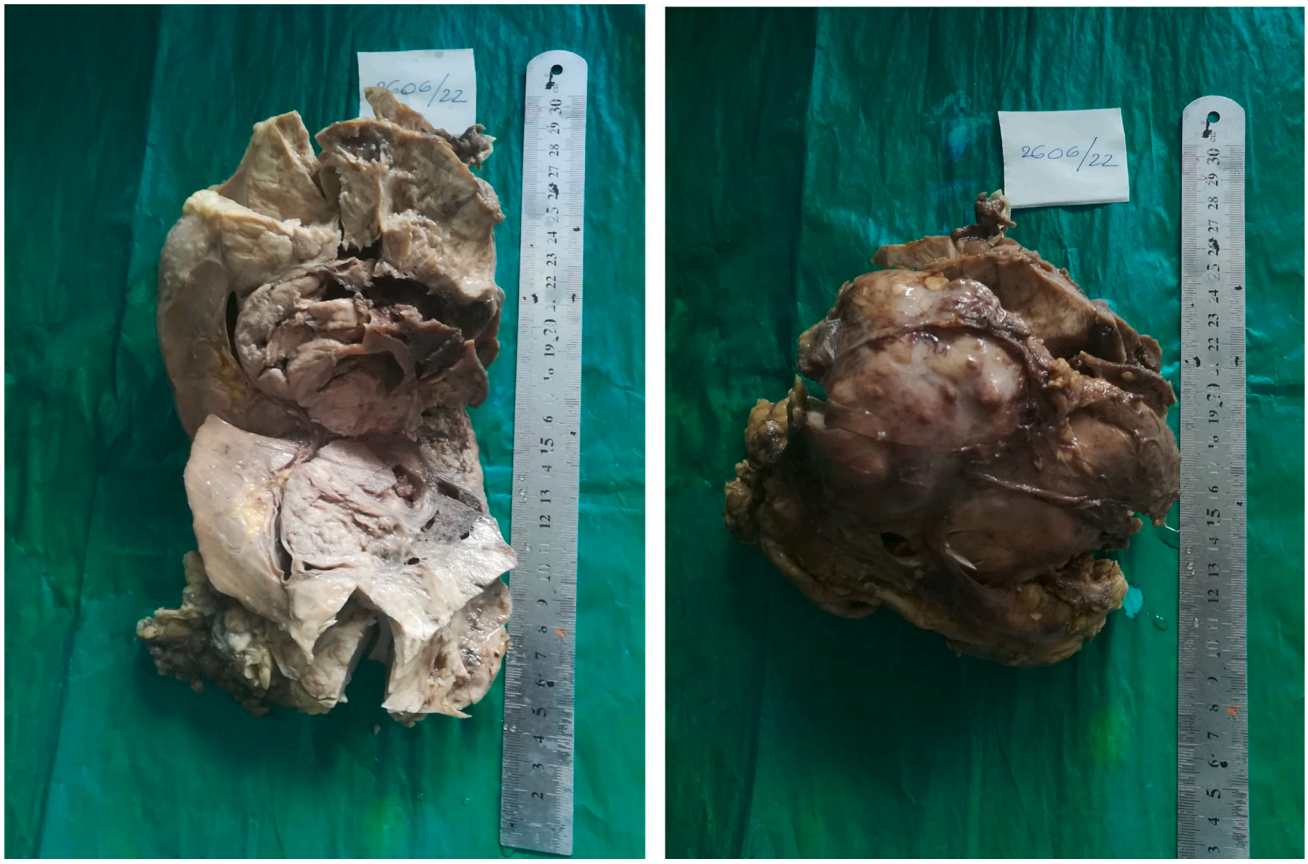


**Fig. 2.** Resected right renal mass.

hematuria and chest related symptoms. There was no family history of renal cancer. Upon abdominal examination, no significant findings were noted. The full blood count report displayed a white cell count of 10.9 $\times$ $10^9$/L, hemoglobin level of 10.5 g/dL, and a platelet count of 292 $\times$ $10^9$/L. The urine full report indicated occasional pus cells and no red cells. Serum creatinine measured 1.13 mg/dL, LDH was 578.5 units, and serum calcium registered 2.2 mmol/L. Abdominal ultrasound revealed a complex solid and cystic mass with mild hydronephrosis on the right side. A chest, abdomen, and pelvis contrast enhanced computed tomography (CECT) scan uncovered a 7 cm right renal mass with a para-aortic lymph node (Staging T3bN1Mx), as well as a 2.3 $\times$ 2.6 cm mass on the upper pole of the left kidney (Staging T1aNxMx) (Fig. 1). Further investigations including 2D echo and transesophageal echocardiography (TOE) showed no atrial extension of the tumor and indicated good left ventricular function. Subsequently, a right radical nephrectomy (Fig. 2) and resection of the para-aortic lymph nodes were performed. The excised tissues were sent for histological analysis. The histopathological examination revealed the presence of malignant spindle cells with necrosis and mitosis, raising suspicion of either primary leiomyosarcoma or unclassified RCC with a pure sarcomatous component. Immunohistochemical analysis confirmed the diagnosis of leiomyosarcoma originating from the right kidney (Fig. 3). The report for the para-aortic lymph node indicated it was reactive.

A follow-up CT scan was performed after a 2-month interval, which

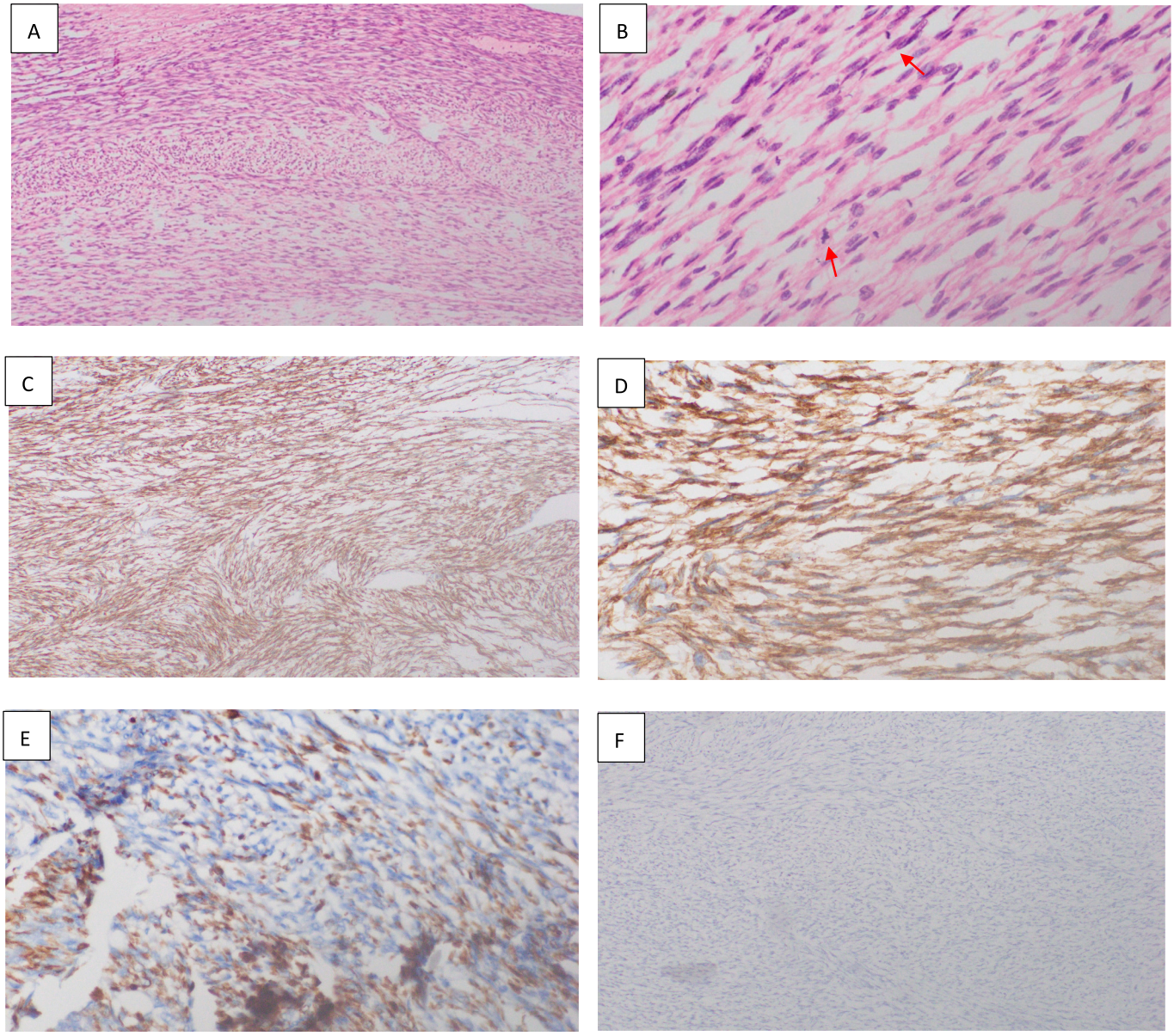


**Fig. 3.** Microscopic images of right renal tissue. (A) - 40× magnification and (B) - 100× magnification H&E stain images, showing leiomyosarcoma which shows fascicles of spindle-shaped cells with elongated cigar-shaped nuclei and mitotic figure (Arrows in the image shows mitotic figures). (C) and (D) Immunohistochemistry smooth muscle actin (SMA) images displaying strong, diffuse staining of tumor cells. (E) Immunohistochemistry with desmin reveals positive staining in tumor cells. (F) S100 stain image shows a negative result in tumor cells.

showed that the left upper pole tumor (2.3 × 2.6 cm) remained stable. Additionally, two lesions were identified in segments IV and VIII of the liver, both measuring less than 1 cm in size (Fig. 4). In response, biopsies were taken from the left kidney and liver, and microwave ablation of the left kidney was performed. The histology of the left renal tissue indicated clear cell renal carcinoma with WHO/ISUP nuclear grade 1. The liver histology demonstrated malignant spindle cell lesion displaying fascicles of undifferentiated spindle cells with irregular hyperchromatic nuclei and frequent mitosis, suggestive of a high-grade undifferentiated sarcoma. Given the context of the patient's history, with a sarcomatous lesion in the right kidney and clear cell renal carcinoma in the left kidney, it was suspected that the liver lesions might represent metastatic deposits from the sarcomatous component of the right kidney (Fig. 5). The case was presented in a multidisciplinary team (MDT) meeting and the decision was made to initiate chemotherapy. However, a follow-up CT scan conducted after 6 weeks revealed a positive response in the renal tumor but the emergence of more new metastases in the liver and peritoneum (Fig. 6).

## 3. Discussion

Renal cancer is considered a relatively common type of cancer, with clear cell carcinoma being a frequently diagnosed histological subtype [1,7,8]. On the other hand, primary leiomyosarcoma is an exceptionally rare occurrence, comprising only 0.5–1 % of all invasive renal tumors. The diagnosis of leiomyosarcoma is established through histological examination, as it lacks distinctive clinical or radiological characteristics [3].

Renal leiomyosarcoma is recognized for its aggressive behavior, characterized by rapid growth, frequent metastases, and a high incidence of both local and systemic recurrences. This aggressive nature contributes to a poor prognosis for patients diagnosed with this condition [9,10].

Leiomyosarcomas exhibit common symptoms as like renal cell

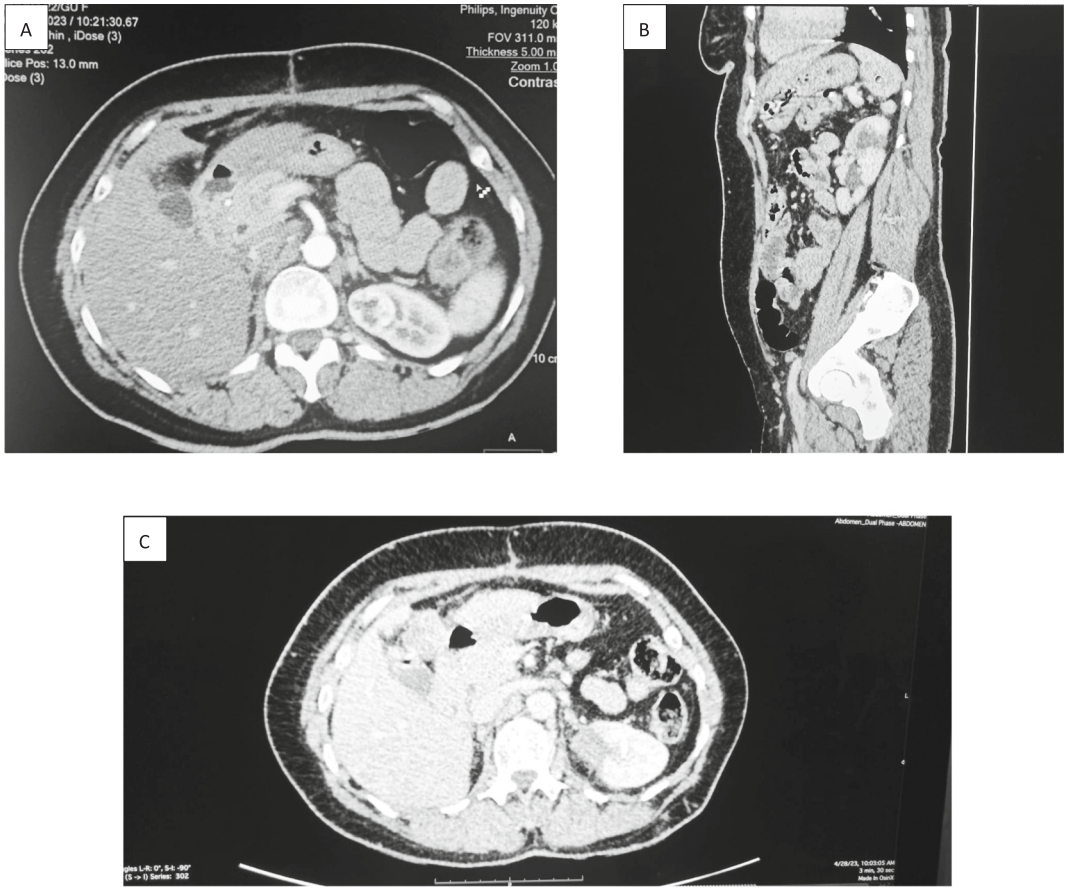


**Fig. 4.** (A) CT image after resection of right renal mass. (B) and (C): CT images showing left renal mass with liver lesion.

carcinoma, such as pain, palpable mass, and hematuria [3]. In the case at hand, the patient's initial presentation was characterized by loin pain, and subsequent assessment using imaging techniques revealed the presence of renal masses.

Localized renal cell cancers are managed through approaches such as partial or radical nephrectomy, ablation, or active surveillance. The choice of treatment method depends on the specific characteristics and features of the identified renal mass [2]. Conversely, for renal sarcomas, the mainstay treatment is radical nephrectomy. However, due to the aggressive nature of these tumors, a combination of triple therapy involving surgery, chemotherapy, and radiotherapy is often recommended [3].

In this specific case, the patient underwent a radical nephrectomy on the right side due to the presence of a large renal mass. The excised sample was subsequently sent to the histology department for further examination.

Dealing with synchronous bilateral renal cancers presents significant challenges in terms of surgical management [5]. When renal cancer is confined to the organ and the contralateral kidney is normal, radical nephrectomy is generally considered the "gold standard" treatment [5]. However, managing bilateral synchronous renal solid masses becomes complex, as standard surgical approaches might lead to insufficient renal tissue for adequate kidney function without the need for dialysis [5].

In the index case of synchronous bilateral renal cancers where each represent different histology, the objective was to perform nephron-sparing surgery, which involves removing the primary tumors while preserving enough renal mass to avoid the necessity of hemodialysis. As a result, a right nephrectomy was carried out while leaving the left renal mass intact. Subsequent follow-up CT scans indicated stability in the left renal mass, but metastatic deposits were identified in the liver. In order to maintain enough viable renal tissue for the patient's overall well-being, microwave ablation of the left kidney was performed.

Histological analysis of the left renal mass indicated the presence of clear cell renal cell carcinoma (ccRCC). The synchronous diagnosis of bilateral renal cell carcinomas (RCCs) is relatively rare, occurring in only 1–5 % of cases [4,11]. According to a case report, bilateral clear cell renal cell carcinomas constituted the predominant histological subtype at 73 %, followed by bilateral papillary renal cell carcinoma at 16 %. Bilateral chromophobe RCC was the third most common type at 4 %. The remaining cases accounted for various combinations of different histological subtypes of renal cancers [12].

According to a report, approximately 30 % of patients diagnosed with ccRCC in localized stages eventually experience the development of metastases, despite undergoing nephrectomy with the intent of a curative approach [2]. Additionally, renal leiomyosarcomas tend to commonly metastasize to the liver, with other potential sites of metastasis including the lung, bone, and stomach [10]. The histopathological examination of the liver revealed a high-grade undifferentiated sarcoma. Considering the patient's history, this finding suggests the possibility that the liver lesion could be a metastatic deposit originating from the sarcomatous component of the right renal cancer.

Despite the implementation of a highly aggressive tumor management approach, a subsequent follow-up CT scan performed after 6 weeks showed a positive response in the renal tumor. However, concerning developments included the appearance of new metastases in both the liver and peritoneum. Patient was transferred to specialized care for continuation of chemotherapy and follow up.

In case of cancer specific survival rates, patients with bilateral renal cancers tended to have lower 5 year and 10 year cancer specific survivals when compared to that of unilateral renal cancer [5]. Due to the rarity of bilateral renal cell cancers, there is a lack of comprehensive

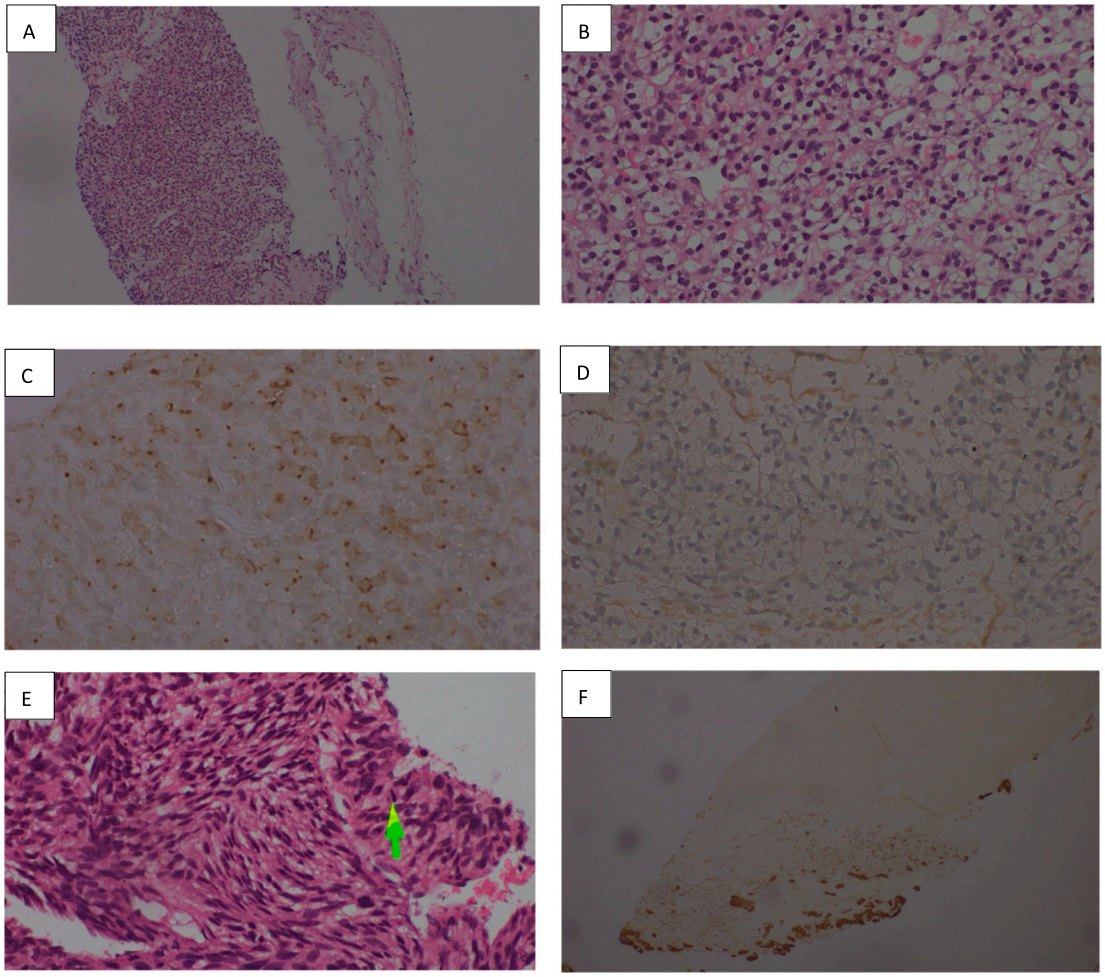


**Fig. 5.** Microscopic images of left renal tissue (A, B, C and D). (A) H&E stain at 40× magnification (B) H&E stain at 400× magnification. (A) and (B) display sheets of cells with clear cytoplasm, distinct cell membranes and intervening network of thin-walled vessels. (C) Immunohistochemistry at 400× magnification with CD10 shows cytoplasmic and membrane positivity in tumor cells. (D) Immunohistochemistry at 100× magnification with SMA indicates a negative result in tumor cells. Microscopic images of the liver biopsy (E and F). (E) H&E stain at 400× magnification, displaying fascicles of undifferentiated spindle cells with irregular hyperchromatic nuclei and frequent mitosis. (F) Immunohistochemistry with pan CK shows a negative result.

management guidelines, and only a limited number of studies have been reported. Moreover, alternative modalities of management, such as radio-ablation and selective tumor embolization, are also limited. Therefore, further studies on the management of bilateral renal cancers are needed to enhance our understanding of its management.

## 4. Conclusion

Bilateral synchronous renal cancers are rare and among them, occurrences of different histologies in contralateral kidneys are even rarer. In case of bilateral synchronous renal cancers, managing with preserving renal mass without the need of dialysis poses a challenge. The prognosis of bilateral synchronous renal cancer is poor, although aggressive and early management is necessary. Given the rarity of bilateral renal cell cancer, there is a lack of comprehensive management guidelines, and only a limited number of studies have been reported. Therefore, further studies on the management of bilateral renal cancer are needed to enhance our understanding of its management.

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## Ethical approval

N/A

## Statement of informed consent

Informed written consent was obtained from the patient in this case study for publication of this case report and relevant images.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Guarantor

Thaka M. Musthafa.

## CRediT authorship contribution statement

Study concept – Thaha M. Musthafa, N. Weerasinghe and Balagobi B. Data collection – Anton Jenil, Munasinghe M.A.D.N and

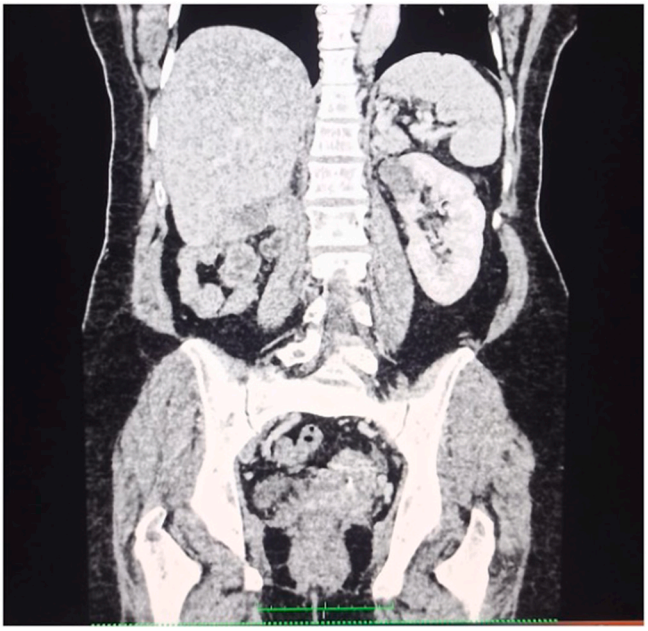

**Fig. 6.** CT image taken after the ablation of left renal tumor.

Priyatharsan K.

Data analysis – Thaha M. Musthafa and Priyatharsan K.

## Declaration of competing interest

None.

## References


[1] J. Hsieh, M. Purdue, S. Signoretti, et al., Renal cell carcinoma, Nat. Rev. Dis. Primers. 3 (2017) 17009, https://doi.org/10.1038/nrdp.2017.9.

[2] J.J. Hsieh, M.P. Purdue, S. Signoretti, C. Swanton, L. Albiges, M. Schmidinger, D. Y. Heng, J. Larkin, V. Ficarra, Renal cell carcinoma, Nat Rev Dis Primers. 3 (2017), 17009. doi: 10.1038/nrdp.2017.9. PMID: 28276433; PMCID: PMC5936048.

[3] S. Dhawan, P. Chopra, S. Dhawan, Primary renal leiomyosarcoma: a diagnostic challenge, Urol Ann. 4 (1) (2012) 48–50, doi: 10.4103/0974-7796.91623. PMID: 22346103; PMCID: PMC3271452.

[4] R. Shigehisa, T. Karashima, S. Kobayashi, et al., Synchronous bilateral renal cell carcinomas with differing histologies, IJU Case Rep. 3 (2020) 196–199.

[5] M.L. Blute, C.L. Amling, S.C. Bryant, H. Zincke, Management and extended outcome of patients with synchronous bilateral solid renal neoplasms in the absence of von Hippel-Lindau disease, Mayo Clin. Proc. 75 (10) (2000 Oct) 1020–1026, https://doi.org/10.4065/75.10.1020. 11040850.

[6] R.A. Agha, T. Franchi, C. Sohrab, G. Mathew, A. Kirwan, A. Thomas, et al., The SCARE 2020 guideline: updating consensus surgical case report (SCARE) guidelines, Int. J. Surg. 84 (1) (2020) 226–230.

[7] P. Jiang, C. Wang, S. Chen, et al., Primary renal squamous cell carcinoma mimicking the renal cyst: a case report and review of the recent literature, BMC Urol. 15 (2015) 69, https://doi.org/10.1186/s12894-015-0064-z.

[8] Bahadoram S, Davoodi M, Hassanzadeh S, Bahadoram M, Barahman M, Mafakher L. Renal cell carcinoma: an overview of the epidemiology, diagnosis, and treatment. G Ital Nefrol. 2022;39(3):2022-vol3. PMID: 35819037.

[9] D. Darlington, F.S. Anitha, Atypical Presentation of renal leiomyosarcoma: a case report, Cureus. 11 (8) (2019) e5433. doi: 10.7759/cureus.5433. PMID: 31632881; PMCID: PMC6797020.

[10] V. Narula, F. Siraj, A. Bansal, Renal leiomyosarcoma with soft tissue metastasis: an unusual presentation, Can Urol Assoc J. \ (3–4) (2015). E139-41. doi: 10.5489/cuaj.2396. PMID: 25844101; PMCID: PMC4375001.

[11] J. Ni, N. Cui, Y. Wang, J. Liu, Case report: bilateral renal cell carcinoma with different histological and morphological features, clear cell and cystic thyroid-like follicular subtype, Front Oncol. 11 (2021), 659706. doi: 10.3389/fonc.2021.659706. PMID: 33981609; PMCID: PMC8107718.

[12] T. Klatte, H. Wunderlich, J.J. Patard, M.D. Kleid, J.S. Lam, K. Junker, J. Schubert, M. Böhm, E.P. Allhoff, F.F. Kabbinavar, M. Crepel, L. Cindolo, A. De La Taille, J. Tostain, A. Mejean, M. Soulie, L. Bellec, J.C. Bernhard, J.M. Ferriere, C. Pfister, B. Albouy, M. Colombel, A. Zisman, A.S. Belldegrun, A.J. Pantuck, Clinicopathological features and prognosis of synchronous bilateral renal cell carcinoma: an international multicentre experience, BJU Int. 100 (1) (2007) 21–25, doi: 10.1111/j.1464-410X.2007.06877.x. Epub 2007 Apr 13. PMID: 17433034.