CASE REPORT

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Sarcomatoid renal cell carcinoma mimicking a non-functioning kidney with stone: A rare case report

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ABSTRACT

Background: Sarcomatoid differentiation is a rare variant of renal cell carcinoma (RCC) and is associated with a worse prognosis. To date, there is still no reliable method to diagnose sarcomatoid renal cell carcinoma (sRCC) preoperatively. This malignancy is sometimes hidden in the minority cases of the non-neoplastic disease, making it difficult to diagnose and leading to diagnostic pitfalls particularly in limited diagnostic tool settings.

Case presentation: A 72-year-old male presented with mild, dull pain in the left flank for one year. The initial assessment revealed a non-functioning left kidney with severe hydronephrosis and a stone. However, after a simple nephrectomy was done, the pathological examination showed a sarcomatoid variant of RCC. In the following weeks, a recurrent inhomogeneous tumor aggressively raised in the left kidney bed infiltrating the adjacent structures to the abdominal wall. An embolization procedure of the tumor feeding artery was carried out followed by administration of tyrosine kinase inhibitor. However, the patient's condition quickly deteriorated and survived for about one month.

Conclusion: In the non-functional kidney associated with stone, a consideration of potentially malignant lesions as a differential diagnosis is suggested. A rare sRCC with the worst prognosis may also exist in this condition. A thorough preoperative examination must be done carefully to avoid a diagnostic pitfall.

Keywords: nephrolithiasis, renal cancer, renal failure, sarcomatoid variant.
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INTRODUCTION

Sarcomatoid renal cell carcinoma (sRCC) is a rare variant of renal cell carcinoma (RCC) with the worst prognosis from all renal tumors.[1] A recent meta-analysis has found that kidney stone was significantly associated with increased risk RCC with a pooled risk ratio of 1.76.[2] Other studies have also reported 3.4-9.3% of malignancy in the nephrectomy specimen removed due to non-functioning kidney.[3,4] The non-functioning kidney made the diagnosis more challenging since the signs and symptoms are unremarkable.

Due to the aggressive nature of sRCC,[1], its recurrence rate is very high. It may also quickly deteriorate the patient's condition in a matter of months.[5,6] Therefore, the preoperative diagnosis of this malignancy needs to be established so the patient can be treated appropriately.

To date, there is still no reliable method to diagnose sarcomatoid renal cell carcinoma (sRCC) preoperatively.[1] This malignancy is so metimes hidden in the minority cases of the non-neoplastic disease,[4] making it difficult to diagnose and leading to diagnostic pitfalls, particularly in the setting of limited imaging modality. Here, we reported a rare case of sRCC that clinically and radiologically mimics non-functioning kidney with stone to emphasize its potential to a diagnosis pitfall, particularly in limited diagnostic tool settings. As far as our knowledge, there is no previous report about it from Indonesia.

CASE REPORT

A 72-year-old male presented with mild, dull pain in the left flank for one year. There was no palpable mass and no increase in pain during palpation. Complete blood count and urinalysis showed no striking abnormalities. Abdominal ultrasonography (USG) revealed severe left hydronephrosis with hyperechoic lesion and acoustic shadow suggesting a stone (Figure 1). Kidney-ureter-bladder (KUB) radiography and intravenous urography (IVU) revealed an opacity at the left paravertebral line measuring 28 x 40 mm. The contrast could not fill through the pelvicalyceal system of the left kidney (Figure 2). The radiographic diagnosis was left renal pelvic stone with severe hydronephrosis and non-visual kidney, suggesting a total obstruction in the left urinary tract. Subsequently, the patient underwent a left nephrostomy with the product of 600 ml transparent yellow urine initially.

The following daily product of left nephrostomy was low. A creatinine

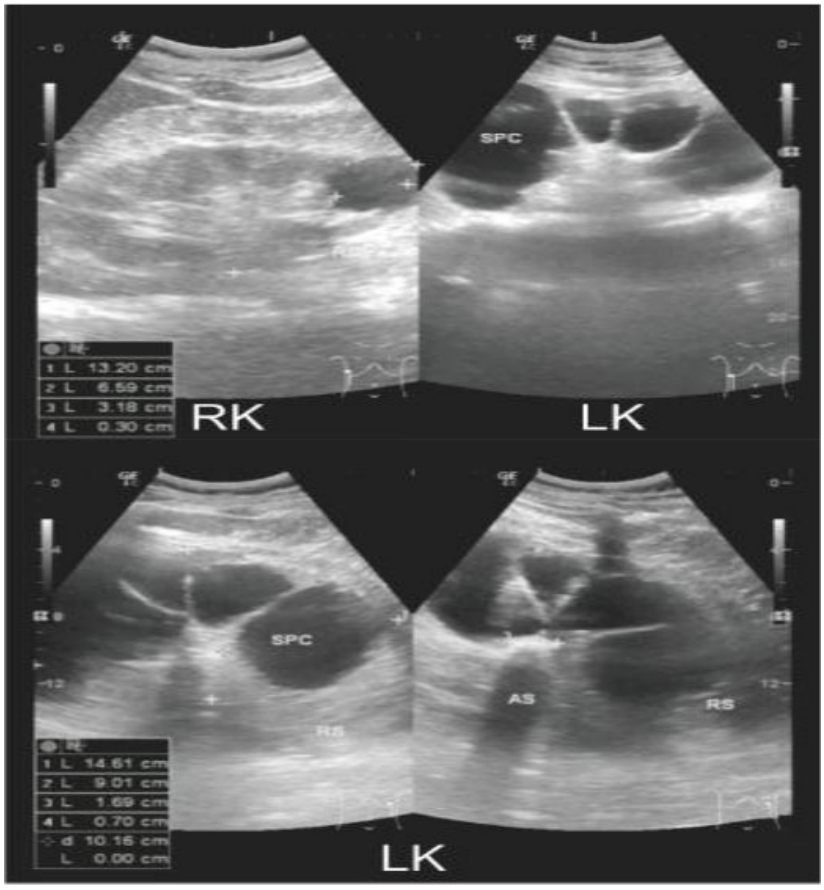

Figure 1. The ultrasound reveals severe left hydronephrosis with thin kidney parenchyma, indicating a poor-functioning left kidney and a hyperechoic lesion with a posterior acoustic shadow suggesting a stone.

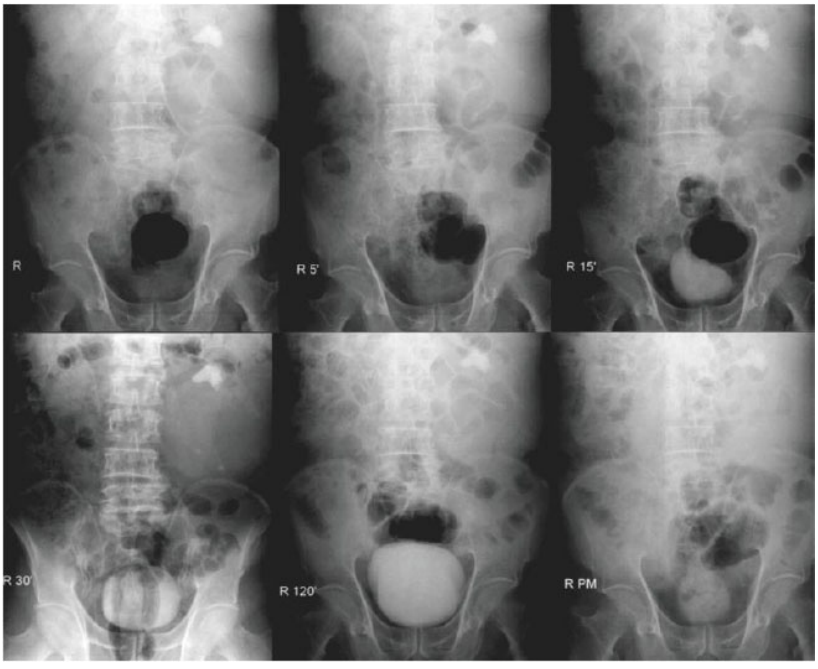

Figure 2. The Kidney-ureter-bladder (KUB) radiography and intravenous urography (IVU) could not reveal the abnormality other than a left kidney stone.

clearance time test performed on the third day for determining the left kidney function revealed 0,71 ml/min. A simple left nephrectomy was done for suspected a non-functioning left kidney with renal pelvic stone. The left kidney was noticeably more significant, with the size of 16x10x9 cm. A complex dark brown colored stone was found inside the kidney, 4 cm in diameter, with a rough, jagged surface obstructing the kidney's pelvic. The patient was in good condition and recovered well postoperatively.

The gross examination of the left nephrectomy specimen showed a kidney with an irregular contour of the external surface, dilated pelvicalyceal system, cortical atrophy, and poorly demarcated yellowish-white mass. Surprisingly, the histopathological examination revealed a cellular tumor arranged in a fascicular pattern consisting mainly of atypical spindle cells. A few tumor cells with clear cytoplasm could still be identified. Morphological findings supported by the positivity of cytokeratin, vimentin, and CD10 on immunohistochemistry confirmed the diagnosis of sRCC (Figure 3). Ki-67 immunohistochemistry staining showed a high proliferative index (57%), indicating aggressive tumor behavior.

Three weeks after nephrectomy, a prominent mass was reappeared in the surgery site (Figure 4), which is poorly identifiable by USG. The following magnetic resonance imaging suggested a recurrent inhomogeneous tumor-infiltrating the adjacent structures to the abdominal wall (Figure 5). An embolization procedure of the tumor feeding artery originating from abdominal aortic branches was carried out to stop the mass growth and control the pain (Figure 6). Therapy continued with administering tyrosine kinase inhibitor, Pazopanib, 800 mg daily, to inhibit tumor angiogenesis. Due to financial reasons, the patient discontinued the treatment and was brought to his home. His family member reported that he was treated for supportive and palliative care by the local general practitioner. The patient survived for about one month.

In this case report, all data were collected from existing clinical findings and diagnostic test results in the medical record. No identifiable information was

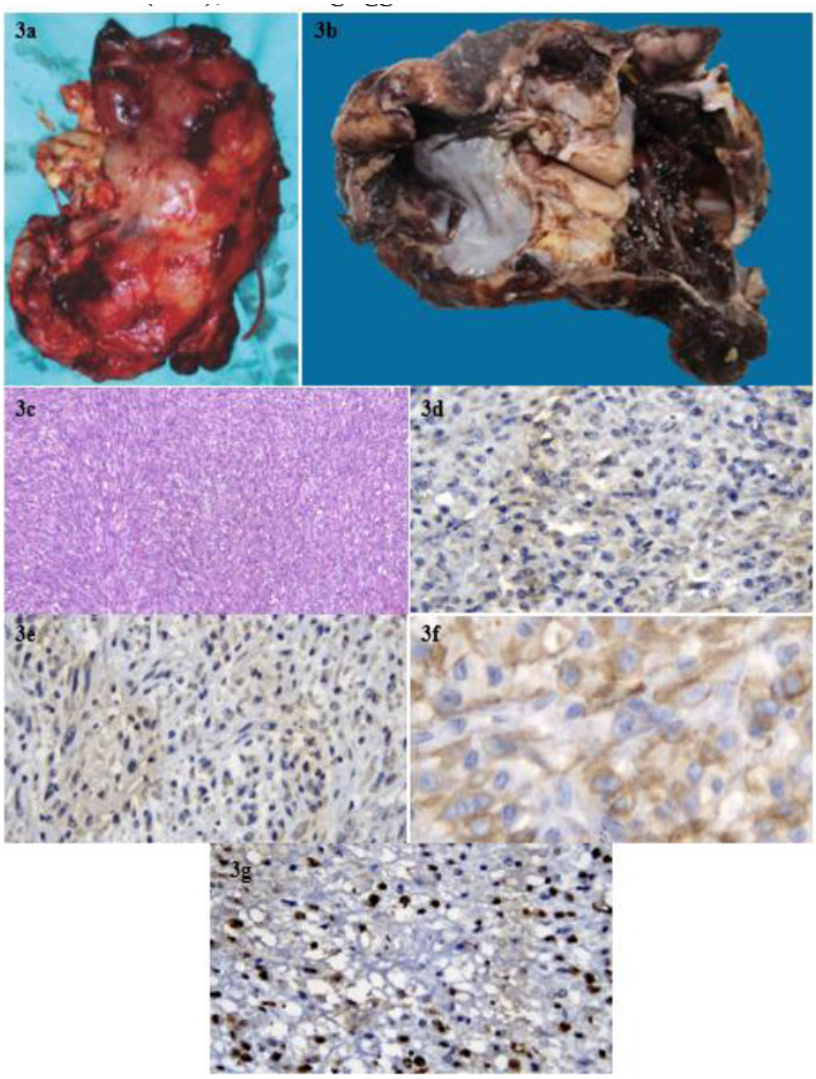

Figure 3. **3a:** Left nephrectomy specimen shows the irregular contour of the outer surface. **3b:** Gross examination reveals pelvicalyceal dilatation and poorly defined yellowish-white mass. **3c:** Microscopic examination shows cellular tumor with fascicular pattern consists of particularly atypical spindle cells. **3d and 3e:** Cytokeratin and vimentin are weakly expressed by some tumor cells. **3f:** Most of the tumor cells strongly express CD10. **3g:** Ki-67 immunostaining shows a proliferative index of 57%.

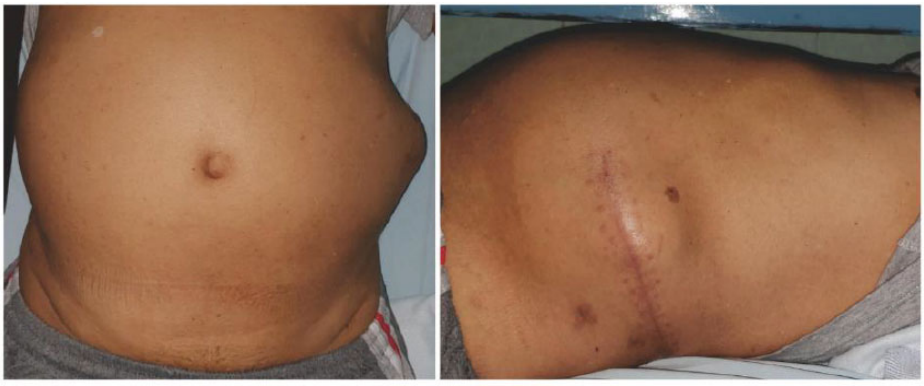

Figure 4. Prominent mass reappeared in the surgical site.

included in this report. The patient had given his consent preoperatively to the usage of the tissue samples for future study purposes. This report protocol was reviewed and approved by the Medical and Health Research Ethics Committee of Faculty of Medicine, Public Health, and Nursing, Universitas Gadjah Mada, with the letter-number KE/FK/1126/EC/2021.

DISCUSSION

To date, there is still no reliable method to diagnose sRCC preoperatively.[1] This malignant kidney lesion was more challenging to diagnose if it is insidious in a non-functioning kidney with thin parenchyma. The chosen imaging modality was IVU that intended to reveal the anatomical and functional abnormality in the severely hydronephrotic kidney. However, the non-functioning kidney could not show the contrast flow in the presented case and revealed no abnormality other than the left nephrolithiasis. On the other hand, a non-contrast-enhanced computed tomography (NCCT) may have a chance to reveal an abnormality in the kidney parenchyma that can be a clue of a malignancy. The NCCT has become the standard in diagnosing kidney stones, replacing the IVU with superiority for determining the stone density, the inner structure of the stone, skin-to-stone distance, and surrounding anatomy, yet, the loss of information on renal function should have been noted.[7]

The severe hydronephrotic left kidney with thin parenchyma revealed from USG and no visible contrast on the IVU leads to the presumption of a non-functioning kidney. Nephrostomy placement under local anesthesia had done to achieve decompression and external drainage of the kidney and followed by determining kidney function by calculating the creatinine clearance time test using the 24-hour urine from nephrostomy. In the setting of malignancy, the nephrostomy placement should be considering the possibility of tumor seeding in the nephrostomy tract.[8,9] Then, the procedure should be indicated for the most emergency condition. Another crucial option in determining kidney function is scintigraphy, which can provide noninvasive information about

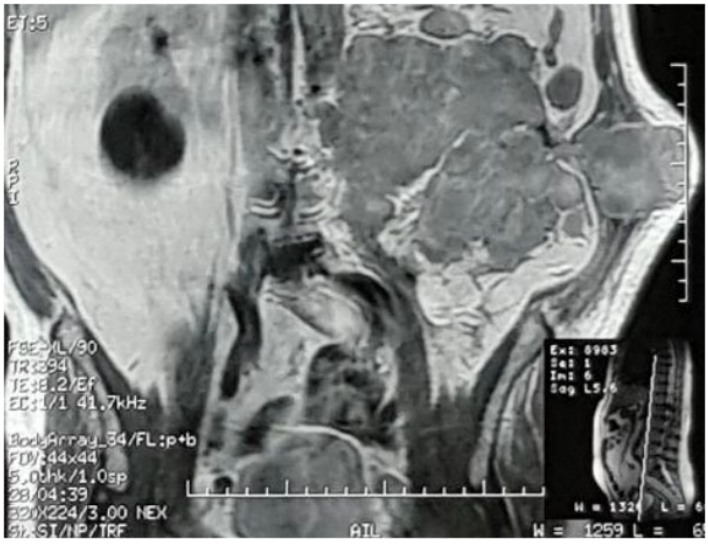

Figure 5. The magnetic resonance imaging shows the recurrent mass infiltrating the abdominal wall and the adjacent structures.

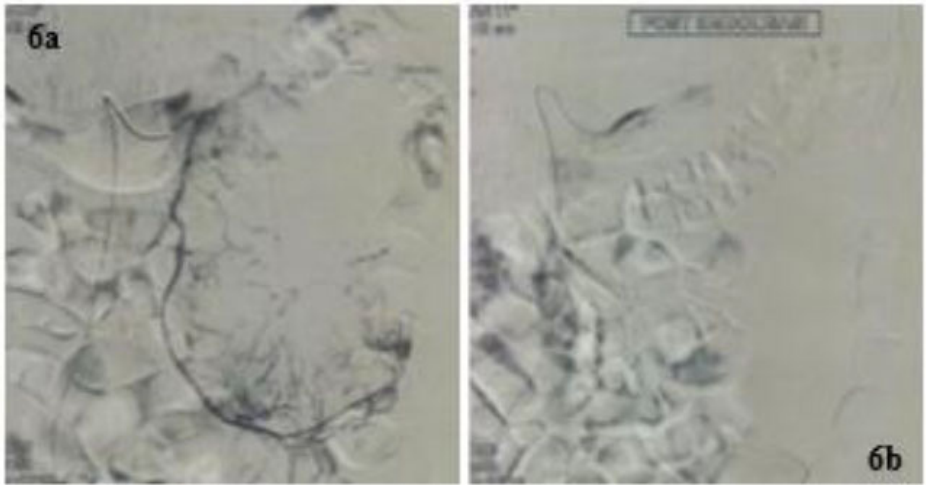

Figure 6. **6a:** During the embolization procedure, the tumor feeding artery noticeably originates from the abdominal aortic branch**. 6b:** The tumor feeding artery is significantly reduced after embolization.

kidney function.[10] Still, we are facing this limitation that scintigraphy is not available in our center.

The non-functioning kidney with stone was treated with a simple nephrectomy. However, in our case, the aggressive local recurrence was raised in a short time afterward due to incidental findings of malignancy. This local recurrence in the kidney bed after nephrectomy is associated with worse survival. It may be due to the incomplete resection of the primary tumor because the simple nephrectomy was performed instead of radical nephrectomy and the aggressive nature of the sarcomatoid variant itself. Several approaches have been suggested to manage local recurrences, such as surgical excision, radiotherapy, systemic treatment, and observation. However, the most optimal local treatment for local recurrence is still under debate due to insufficient evidence.[11] In our case, we consulted the interventional radiologist to perform the embolization to control the pain and reduce the mass by plugging the tumor's feeding artery. According to European Urology guidelines, embolization may control symptoms for patients unfit for surgery or with a non-resectable disease.[11]

The risk of RCC associated with kidney stones was recently studied in a meta-analysis, with a significant pooled risk ratio of 1.76 (95% CI 1.24-2.49). The association of renal stone with urothelial carcinoma is higher, with a pooled risk ratio of 2.14 (95% CI, 1.35-3.40).[2] In addition to these findings, the presence of stones in a non-functioning kidney should also become a concern because of the possibility of malignancy. Other studies reported that the incidence of malignancy was 3.4 and 9.3% of all nephrectomy upon the non-functioning kidney, and non-functioning kidney associated with stone, respectively.[3,4] It proposed that chronic irritation or infection related to stone may play a role in cancer development.[12] Regarding these findings, clinicians should consider the possibility of a malignant condition until it is proven otherwise.

Although it accounts for 5 percent of RCC, the sRCC remains poorly treatable and highly lethal kidney cancer. It is associated with more aggressive disease and poor outcomes after nephrectomy with a high recurrence rate. Because of its rarity, dedicated trials may not be easy, and the treatment remains failed to meet the goals.[1,5,13]

In the non-functional kidney associated with stone, a consideration of potentially malignant lesion as a differential diagnosis has been suggested. A rare sRCC with the worst prognosis may also exist in this condition. The management approach of the kidney with suspected malignancy will be different with a non-functioning kidney with stone. The diagnostic imaging may be challenging because contrast cannot flow through the kidney and enhance the malignant lesion. A thorough preoperative examination must be done carefully to avoid a diagnostic pitfall.

CONCLUSION

Sarcomatoid renal cell carcinoma (sRCC) is a rare variant of renal cell carcinoma (RCC) with the worst prognosis from all renal tumors. However, after a simple nephrectomy was done, the pathological examination showed a sarcomatoid variant of RCC. In the following weeks, a prominent and aggressive mass was raised in the previous surgical site. Although extremely rare, the surgeons should

consider sRCC as the differential diagnosis for a case of renal stone.

FUNDING

This research did not receive any funding.

ETHICAL APPROVAL

This report protocol was reviewed and approved by the Medical and Health Research Ethics Committee of Faculty of Medicine, Public Health, and Nursing, Universitas Gadjah Mada, with the letter-number KE/FK/1126/EC/2021.

CONSENT

Written informed consent for publication of this case report with accompanying images was obtained from the patient.

AUTHOR CONTRIBUTION

All authors contributed to the collection of patient data and writing process of the manuscript. Faridz Albam Wiseso: Conceptualization; Data curation; Investigation; Methodology; Resources; Validation; Visualization; Writing; Supervision. Raden Danarto: Conceptualization; Data curation; Investigation; Methodology; Resources; Validation; Visualization; Writing; Supervision. Hanggoro Tri Rinonce: Resources; Data curation; Validation; Visualization; Writing; Supervision. Dewa Nyoman Murti Adyaksa: Conceptualization; Writing. Yana Supriatna: Conceptualization; Writing.

CONFLICT OF INTEREST STATEMENT

nil.

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