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CASE REPORT

A Rare Case of Sarcomatoid Renal Cell Carcinoma in a Young Adult Patient

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Background: Sarcomatoid differentiation is a rare condition that could present in different subtypes of renal cell carcinomas (RCCs) and is associated with a significantly poor prognosis. Sarcomatoid renal cell carcinoma (SRCC) patients are typically aged between 54 and 63, with a male-to-female ratio ranging from 1.3:1 to 2:1. Here, we report a case of SRCC in a 29-year-old female patient.

Case Presentation: A 29-year-old female presented with left flank pain. A large lump was palpated on left flank and there was costovertebral angle tenderness. The lump was enlarged, and the patient also suffered from anemia. Abdominopelvic CT demonstrated solid mass with an internal gliosis in the left part of the renal cortex and the solid component was enhanced with contrast admission. Then, the patient underwent left radical nephrectomy with wide perirenal excision and paraaortic lymph nodes resection. Histopathological examination revealed SRCC with no lymphovascular invasion.

Conclusion: The scarcity of data on SRCCs emphasizes the need for ongoing research into the biology, diagnostics, and effective treatment options for patients with this disease, as responses to conventional therapies have been disappointing, leaving patients with few options. Cytoreductive nephrectomy for SRCC patients with metastatic disease is debatable, although some research suggests resection at any stage in patients with good performance status. In this case, radical nephrectomy was performed and there was no evidence of metastasis.

Keywords: flank, poor prognosis, renal cell carcinoma, sarcomatoid differentiation

Introduction

Renal cell carcinoma (RCC) is a biologically diverse group of cancers. A new histological classification has differentiated the more prevalent clear cell RCC (ccRCC) (about 80% of all RCC cases) from the other subtypes, collectively known as nonclear cell RCC (nccRCC). Any RCC histology, both ccRCC and nccRCC, may be associated with occurrence of sarcomatoid differentiation.[1,2] The incidence of sarcomatoid transformation varies by RCC subtype, but is higher in clear cell (5–8%) and chromophobe (8–10%) RCCs.[3] Approximately 20% of metastatic RCC cases have sarcomatoid differentiation. Sarcomatoid renal cell carcinoma (SRCC) patients are typically aged between 54 and 63, with a male-to-female ratio ranging from 1.3:1 to 2:1. Here, we report a case of SRCC in a 29-year-old female patient, which is rare in this age group.

Case Presentation

A 29-year-old female presented with left flank pain for 4 months before admission. There was no hematuria. A large lump was palpated on left flank and there was costovertebral angle tenderness. Laboratory examination revealed low hemoglobin (Hb = 8.8 g/dL), hematocrit (28.1%), red blood cell count (3.68 million/µL), MCV (76.4 fl), MCH (23.9 pg), and MCHC (31.3 g/dL). There was an increased platelet and erythrocyte sedimentation rate (ESR). Urinalysis revealed high bacterial count. Abdominopelvic CT demonstrated both kidneys in normal shape and size. The cortical and

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medullary boundaries were clear. A dense mass (HU 40) with surrounding gliosis (HU 20) was seen in the left part of the renal cortex, which was well circumscribed and regular. After contrast administration, an increase in the solid component (HU 60) was seen. The size of the lesion was approximately 7.5 × 11.4 × 11.8 cm (Figure 1). The patient received blood transfusion for anemia correction. But it was later found that the lump was enlarged and the patient also complained about frequent dizziness and dark vision.

The patient underwent left radical nephrectomy with wide perirenal excision and paraaortic lymph nodes resection. Macroscopic examination showed that the kidney looked lumpy, brownish gray in color, and cystic, with a size of 12 × 5 × 4 cm. In the cystic part, the size was 13 × 9 ×13 cm. On lamellar cutting, there were cystic areas with brownish red liquid and the inner surface was a necrotic, brittle, and hemorrhagic mass. The ureter was 5 cm long and 0.5 cm in diameter. Microscopic examination from necrotic and hemorrhagic masses tissue revealed lysed cells with minimal bleeding and infiltration of inflammatory cells (Figure 2A). Tissue preparation from the periphery of the lysis area revealed a loose tissue with clusters of polymorphic, round, oval, spindle, stellate cells with enlarged nuclei, vesicular, and difficult to see cytoplasm (Figure 2B). Multinucleated giant cells and rhabdoid cells were also found. No lympho-vascular invasion was found. Based on these findings, it was concluded that it was sarcomatoid renal cell carcinoma.

Macroscopic examination of the excised lymph nodes revealed that they were gray in color, chewy, and 1 cm in diameter. Microscopic examination revealed a proliferation of secondary lymphoid follicles in the cortex that extended to the paracortex. Around the follicle there was bleeding accompanied by a light neutrophil infiltration (Figure 3). No signs of malignancy were found. And it was concluded that it was reactive follicular hyperplasia. Based on these findings, the patient was diagnosed with SRCC with clear cell carcinoma (grade IV) as a differential diagnosis.

Three-months post-surgery, the patient had no complaints and was in good condition; laboratory examination showed low LDH (128 U/L) in blood. In urine it was found that patient has high leucocyte esterase (500 cell/microliter), high leucocyte (294.2/microliter), and high bacterial count (3121.1/microliter). Other blood examination and urinalysis were within normal limits. Abdominal CT scan showed that neither images of mass nor enlargement of lymph node were found (Figure 4).

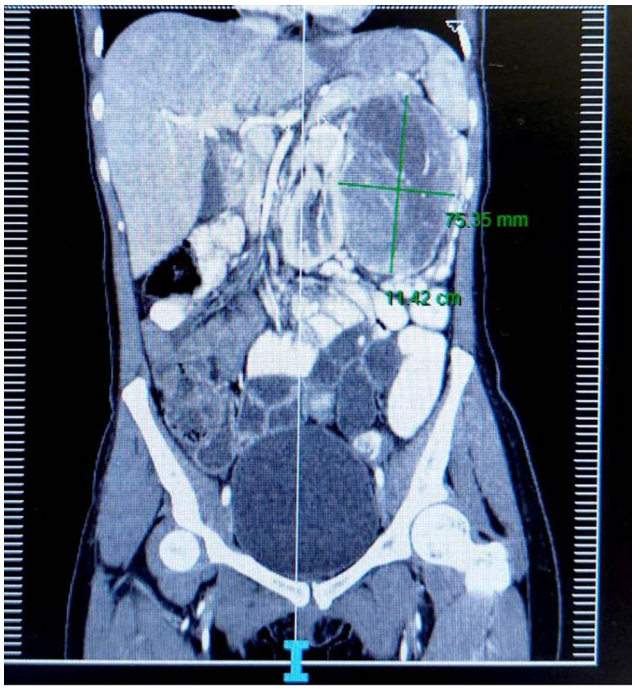

Figure 1 Abdominopelvic CT scan with contrast demonstrated solid mass with an internal gliosis in the left renal cortex. The solid component is enhanced with contrast admission, suggestive of malignancy.

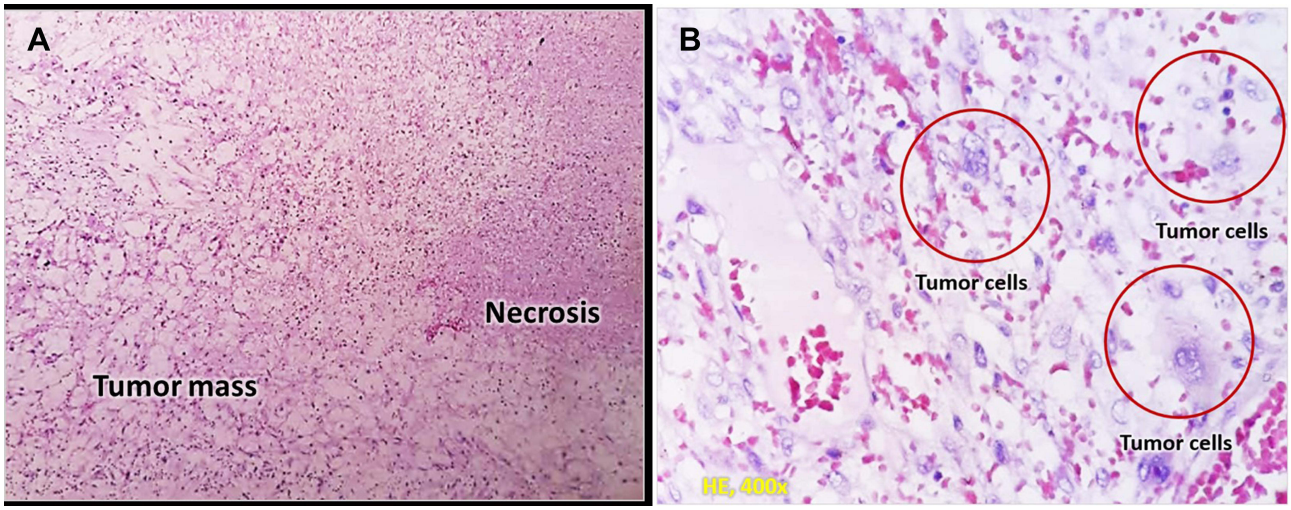

Figure 2 (**A**) Microscopic examination of renal tissue revealed tumor mass and necrosis. (**B**) Microscopic examination from the periphery of the lysis area revealed a cluster of tumor cells.
Abbreviation: HE, hematoxylin and eosin stain.

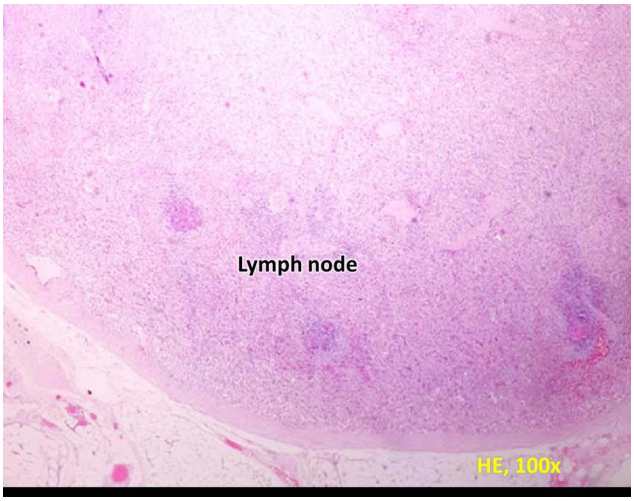

Figure 3 Microscopic examination of the excised lymph nodes.
Abbreviation: HE, hematoxylin and eosin stain.

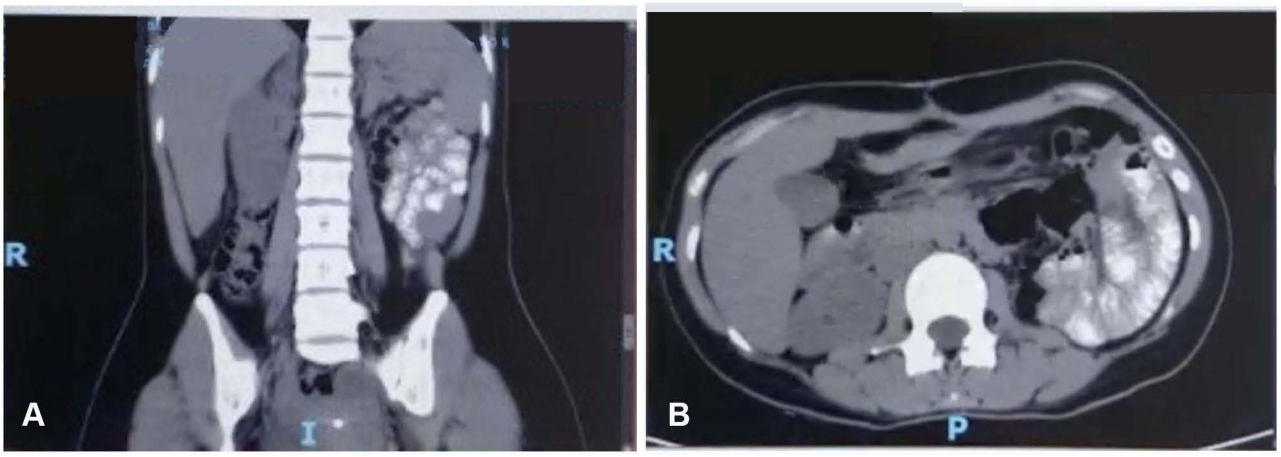

Figure 4 (**A**) Coronal view CT scan in patient three months post-surgery. (**B**) Axial view CT scan in patient three months post-surgery.
Abbreviations: R, right side; P, posterior side.

Discussion

Previously thought to be a renal sarcoma, SRCC is now recognized as a type of dedifferentiated carcinoma and thus no longer a distinct histologic entity. SRCC is a form of RCC in which a portion of the tumor transforms into a high-grade undifferentiated component characterized by the presence of spindle cells, but ultrastructural and immunohistochemical analyses show that the tumors have both epithelial and mesenchymal differentiation.[3–5] SRCC implies an evolution from a differentiated to an undifferentiated tumor characterized by epithelial–mesenchymal transition (EMT) due to a genetic divergence between epithelial and mesenchymal elements from a common cell of origin.[2,6] It is frequently studied as a distinct clinical entity because it is associated with an aggressive phenotype, advanced stage at diagnosis, and poor overall survival (OS) and disease-specific survival (DSS).[7,8] Furthermore, it has a poor response to cytotoxic chemotherapy, cytokines, and targeted therapies.[3,9,10] Notably, having a higher percentage of sarcomatoid features is linked to a lower chance of survival.[9–11] The overall median survival of SRCC was reported to be 4–9 months after diagnosis.[4,12]

Approximately 20% of metastatic RCC cases have sarcomatoid dedifferentiation. Patients with SRCC typically present between the ages of 54 and 63, with a male-to-female ratio ranging from 1.3:1 to 2:1. In this case, the patient is a young adult female, which is not in line with the majority of cases. The effect of gender difference, like those underlying all other RCCs, remains unknown. However, historical effects of gender differences on occupational exposure or smoking status, as well as sex hormones on tumor biology, are all possibilities.[13]

The appearance of SRCCs varies according to severity of the disease at the time of diagnosis. As most of the cases were locally advanced or metastatic, almost all patients were symptomatic at the time of presentation.[13] Typical signs and symptoms include flank or abdominal pain (present in 51% of symptomatic patients), hematuria (22–34.7%), weight loss (18–22.6%), fatigue (15%), fever (6–10.6%), night sweats (6–12.6%), and cough and/or dyspnea (6%).[13] The main sites for distant metastasis were the lungs, bone, lymph nodes, liver and brain.[9,13,14] In this case, there is no sign of metastasis.

SRCCs are frequently large (median 10 cm), usually seen as dense gray or white areas in the tumor architecture and look firm and fleshy when dissected. The presence of epithelial and sarcomatoid components is the typical microscopic finding of SRCCs. Compared to RCCs, the sarcomatoid part lacks recognizable epithelial components and histologically resembles pleomorphic sarcoma with abnormal and hypercellular spindle cells. These sarcomatoid differentiation regions can be either heterogenous or homogenous. Coagulative necrosis is usually observed in almost all SRCC cases while less than 50% have microvascular invasion. About 15% of SRCC patients were observed to have rhabdoid characteristics.[13] A 2016 WHO guideline stated that occurrence of sarcomatoid differentiation is adequate to diagnose RCC due to its distinct features.[1]

Surgical procedure is still the preferred treatment when diagnosed at early stage. However, preoperative imaging or biopsy are often uncertain as most cases are metastatic with systemic symptoms.[13] A retrospective study involving the management of metastatic RCCs showed a better survival in patients who underwent cytoreductive surgery prior to systemic therapy than systemic therapy alone.[15–17] The advancement in immunotherapies for RCCs has allowed for the discovery of exploratory biomarkers that could help in the treatment of SRCCs, although more research is required for a proven effective agent.[13] In this case, it was an early stage tumor and thus the patient received surgery. The patient underwent left radical nephrectomy with wide perirenal excision and paraaortic lymph nodes resection.

SRCC prognostic factors include the scale of sarcomatoid parts, surgical stage, tumor size, and genetic factors. Furthermore, multiple studies have found that tumors with a high TNM (tumor node metastasis) stage are a poor predictor of survival. Another important prognostic factor is tumor necrosis. Tumors with increased necrosis in sarcomatoid areas have a lower survival rate, whereas the amount of histologic tumor necrosis has no effect on outcome. Tumor size may also play a role in prognosis, as it is an independent predictor of cancer-specific mortality.[4]

Conclusion

The scarcity of data on SRCCs emphasizes the need for ongoing research into the biology, diagnostics, and effective treatment options for patients with this disease, as responses to conventional therapies have been disappointing, leaving patients with few options. Cytoreductive nephrectomy for patients with SRCC who have metastatic disease is debatable, but some research suggests resection at any stage in patients with good performance status. In this case, radical nephrectomy was performed and there is no evidence of metastasis.

Ethics Approval

This study was approved by the Ethics Committee of the Faculty of Medicine, Universitas Sumatera Utara with letter number 218/KEP/USU/2022.

Consent for Publication

The patient provided written informed consent for the case details to be published.

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Disclosure

The authors have declared that no competing interests exist in this work.

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