

Renal Pelvis Squamous Cell Carcinoma with Sarcomatoid Transformation-Review of a Rare Case with Clinical and Pathological Findings

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Abstract

Primary renal pelvis Squamous Cell Carcinoma (SCC) with sarcomatoid transformation is an extremely rare neoplastic entity. The present study reports an unusual case of renal SCC associated with sarcomatoid transformation in an 81-year-old female with a history of nephrolithiasis. Abdominal CT study revealed an 11.6 cm cystic mass in the left kidney. Histopathology and immunohistochemical examination of the radical nephrectomy specimen showed squamous cell carcinoma with sarcomatoid transformation in the renal pelvis. To better understand the disease a systematic literature review was performed.

Keywords: Squamous cell carcinoma; Sarcomatoid transformation; Kidney

Introduction

Urothelial cell carcinoma is the most common malignancy in the renal pelvis. Renal pelvis squamous cell carcinoma (SCC) is a particularly rare entity, accounting for approximately 1% of malignant renal neoplasm in the United States according to WHO classification [1,2]. To our knowledge, there are four reported cases in the English medical-related literature in the PubMed archived system. This entity is often associated with chronic infection and irritation, such as long term urolithiasis, smoking, hydronephrosis, toxins, instrumentation and Vitamin A deficiency [2,3]. Squamous metaplasia and urothelial dysplasia of the pelvis are risk factors. The mechanism for the sarcomatoid transformation is not known. Renal pelvis primary SCC has an insidious onset with vague symptoms such as abdominal or flank pain, hematuria, and fever. The radiologic findings are usually non-specific, which results in delayed detection of disease. The prognosis of renal SCC with sarcomatoid transformation is particularly poor.

Case Report

An 81-year-old female with a history of nephrolithiasis for prolonged duration was admitted for a kidney mass and sepsis. Abdominal CT scan revealed an enlarged left kidney with an 11.6 × 9.9 cm mass extending into the renal hilum and a central 7.4 × 4.7 cm complex collection of fluid with gas suggesting emphysematous pyelonephritis. Multiple calculi were identified with dilated calyces in the mid to lower pole (Figure 1). Subsequently, a left radical nephrectomy was performed.

Gross examination of the resected left kidney showed a unifocal, poorly defined mass (11.6 × 7.0 × 4.5 cm) at the mid to lower pole associated with extensive necrosis, hemorrhage, and cystic change. The mass compressed the pelvis producing marked hydronephrosis. The cut surface of the mass displayed a pale-tan, firm, solid, fleshy and variegated appearance (Figure 2).

Microscopic examination revealed a malignant neoplasm displaying typical histological features of SCC with keratinization and high-grade sarcomatoid features involving a portion of the mass. The sarcomatoid area showed a pleomorphic population of spindle cells in a fascicular pattern with >20 mitosis/10 HPF. Vascular invasion and perineural invasion were present. The mass extended into perinephric sinus fat. In the renal pelvis, the epithelial lining showed squamous metaplasia and focal high grade squamous dysplasia. The underlying invasive squamous carcinoma exhibited keratin pearl formation and intercellular bridging. The transition zone from the usual type of SCC to the areas with high-grade sarcomatoid features was abrupt (Figure 3).

Immunohistochemical studies showed the SCC component was positive for pancytokeratin and negative for vimentin, while the sarcomatoid component was positive for vimentin and negative for desmin and S100. P40 was positive in both the SCC and sarcomatoid components. Ki-67 proliferative index was approximately 50% in the sarcomatoid component and approximately 5% in the SCC component. Final pathologic diagnosis was primary renal pelvis SCC with high grade sarcomatoid transformation (Figure 4).

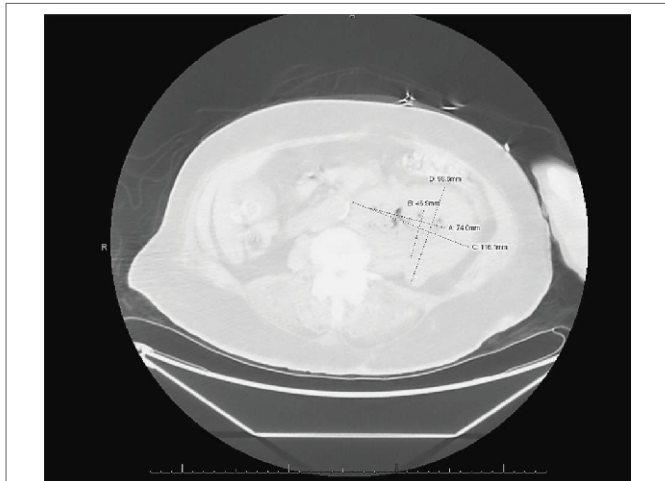


Figure 1: Computerized Tomography (CT) study in the left kidney. CT scan showed an 11.6 cm mass at the mid to lower pole.

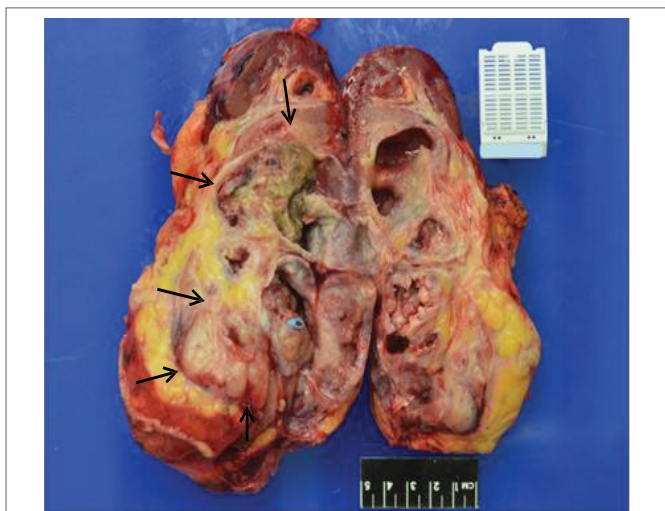


Figure 2: Gross image of the left kidney. A poorly defined kidney mass (11.6 × 7.0 × 4.5 cm) was located at the mid to lower pole. The cut surface was firm and fleshy with marked necrosis, hemorrhage and cystic change.

Results

(Figures 1-4).

Discussion and Conclusion

Primary renal pelvis SCC with sarcomatoid transformation is a very uncommon clinical entity [1,2]. This aggressive tumor arises in the pelvis and is closely associated with chronic nephrolithiasis, infection or irritating factors. Nephrolithiasis is the most common risk factor [3-10]. Due to repeated and chronic irritation of the urothelial surface, the renal pelvic lining may exhibit squamous metaplasia and dysplasia, finally resulting in invasive squamous carcinoma. The adjacent renal parenchyma may be involved as it was in our case. Primary renal SCC is highly aggressive, often at an advanced stage at the time of diagnosis. Sarcomatoid transformation portends a much worse prognosis. Sarcomatoid transformation of renal malignancy was initially described in 1961 [8]. The etiology of the transformation is still under investigation with no proven

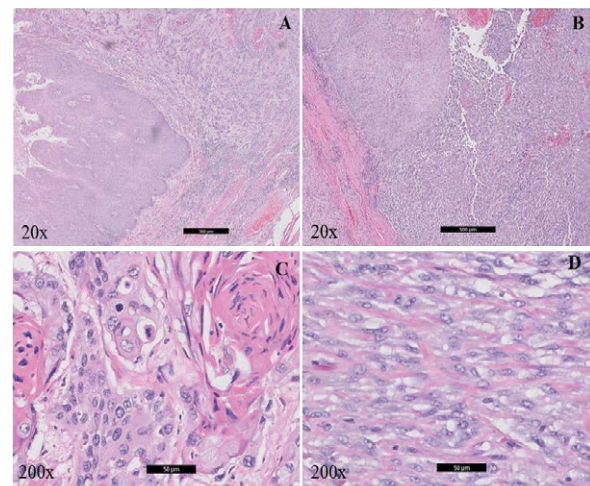


Figure 3: Histopathological findings of the left kidney mass (H&E stain). A) Invasive squamous cell carcinoma with overlying keratinized squamous metaplasia and dysplasia (20X); B) Squamous cell carcinoma with sarcomatoid transformation (20X). C) Invasive squamous cell carcinoma with keratin pearl and mitotic figures (200X); D) Sarcomatoid transformation showing spindle cell morphology with enlarged, irregular nuclei and prominent nucleoli (200X).

molecular pathway identified. The reported cases of renal pelvis SCC with sarcomatoid change all presented with advanced stage disease [9,10].

There are no standard guidelines for the treatment of this cancer entity. Radical nephrectomy is the most commonly pursued treatment option [3-10]. Our patient presented with emergent sepsis and underwent a radical nephrectomy with an uneven clinical course. The surgery revealed rupture of the large necrotic kidney. At the time of diagnosis our patient was already in the advanced stages of disease. Co-existing sepsis complicated the post-surgical recovery. Renal pelvis SCC with sarcomatoid transformation often shows an aggressive course with poor prognosis [1,2,8-10]. This malignancy does not respond well to chemotherapy, radiation therapy or surgery [9,10]. Our patient stabilized and was discharged with post-operative follow up for four weeks. She was re-admitted and subsequently died of disease complications.

Overall, patients with renal pelvis SCC present with more advanced disease and have lower survival rates compared to their urothelial cell carcinoma counterparts. Development of an effective protocol for the treatment of this cancer is a challenge due to the rarity of primary renal pelvis SCC with sarcomatoid transformation and the highly aggressive disease course. Our case may provide additional information to promote early and accurate diagnosis, and optimal disease management.

Ethical Approval

The presented investigations and the publication of data comply with ethical rules of East Carolina University and general rules of experimentation with humans.

Declaration of Conflicting Interests

There are no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

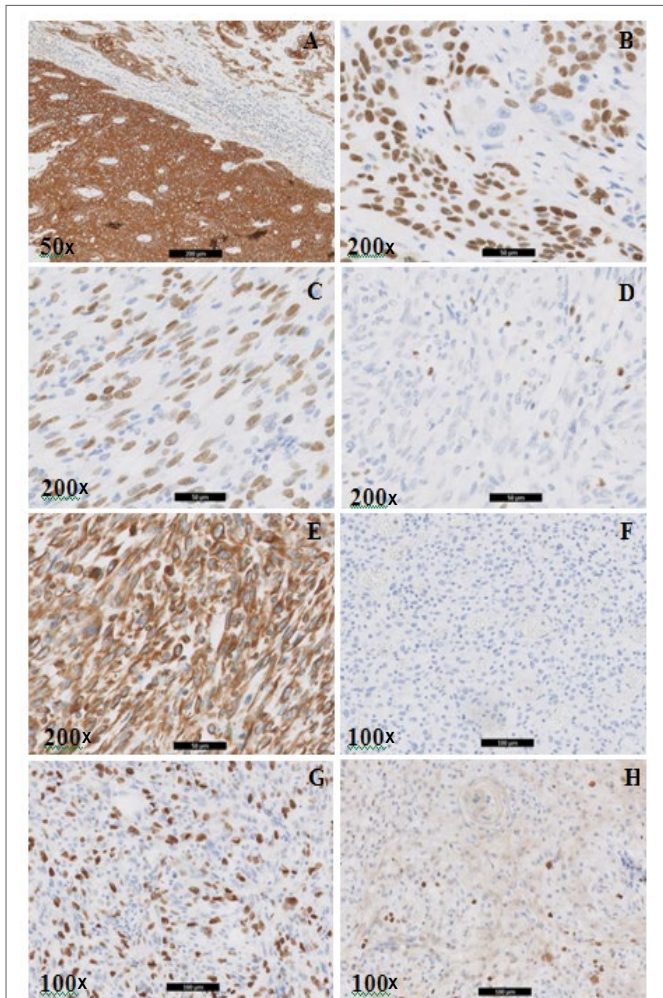


Figure 4: Immunohistochemical studies of the left kidney mass. SCC areas are diffusely and strongly positive for pancytokeratin (A, 50X) and positive for P40 (B, 200X). Sarcomatoid areas are positive for P40 (C, 200X), mostly negative for GATA-3 (D, 200X), diffusely and strongly positive for vimentin (E, 200X) and negative for desmin (F, 100X); Sarcomatoid areas showed a high Ki-67 proliferative index (G, 100X). SCC areas showing a low rate of Ki-67 proliferative index (H, 100X).

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