Squamous cell carcinoma of the renal pelvis masquerading as xanthogranulomatous pyelonephritis

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We report a rare case of a 56-year-old male with a history of hypertension who initially presented to the emergency department with abdominal pain and was radiologically diagnosed with left xanthogranulomatous pyelonephritis (XGP) in a non-functioning kidney with a staghorn calculus. Pathological evaluation of his kidney revealed squamous cell carcinoma (SCC) of the renal pelvis with invasion into the renal parenchyma. We highlight the presentation, diagnosis, and management of this rare condition.

Key Words: renal pelvis tumor, squamous cell carcinoma, xanthogranulomatous pyelonephritis, staghorn calculus

Introduction

Carcinomas of the renal pelvis and ureter account for only about 1% of all urogenital malignancies.[1,2] Within this category, primary squamous cell carcinoma (SCC) of the renal pelvis accounts for only 0.5%-0.8% of malignant renal neoplasms.[1,2] These patients typically lack an identifiable clinical presentation, as they can present without pain, hematuria, or a palpable mass.[1] Epidemiologically, primary renal SCC usually occurs in adults aged 50-70 years old, without sex predominance.[2] These tumors are clinically and radiologically insidious, and thus are often diagnosed when already locally advanced and/or metastatic.[3] Given their often late stage presentation, these tumors tend to be aggressive and invasive with a poor prognosis.[4] Major risk factors associated with SCC of the renal pelvis include chronic renal infection with or without vesicoureteral reflux, nephrolithiasis (especially staghorn calculi), and inflammation, which cause squamous metaplasia that can lead to SCC.[2-4] Other less common risk factors include endogenous and exogenous chemicals, hormonal imbalance, vitamin A deficiency, smoking, and schistosomiasis.[3] Pathology of SCC of the renal pelvis often displays infiltration of the renal parenchyma.[2] We report a complicated case of SCC of the left renal pelvis diagnosed after radical nephrectomy performed for a staghorn calculus in a left non-functional kidney with presumed XGP.

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Case report

The patient is a 56-year-old male with a past medical history of hypertension on valsartan who presented to the emergency department complaining of 3 weeks of abdominal pain radiating to his bilateral flanks, nausea, vomiting, and weight loss. He had known chronic left hydronephrosis with a left staghorn calculus diagnosed years previously. Nephrectomy was recommended by his local urologist at this time but given that he was asymptomatic he elected to defer definitive treatment. On admission he was found to have an acute kidney injury with an elevated creatinine to 2.2 mg/dL (from baseline 0.8) and concern for a urinary tract infection. However, his urine culture ultimately resulted no growth. Computed tomography (CT) of the abdomen and pelvis demonstrated left upper pole severe chronic hydronephrosis with a large staghorn calculus, Figure 1. Additionally, this showed a heterogenous renal mass with dystrophic calcifications occupying the mid and lower poles of the left kidney with expansion outside of Gerota's fascia but not invading nearby organs suspicious for possible renal malignancy. Magnetic resonance imaging (MRI) was obtained to further characterize the lesion which demonstrated a heterogeneous expansile non-enhancing soft tissue lesion of the inferior renal pole with preservation of the renal cortex and focal restriction on diffusion weighted imaging suggestive of XGP rather than renal neoplasm. Follow up nuclear renal scan showed 2% function of the left kidney with an obstructive drainage pattern.

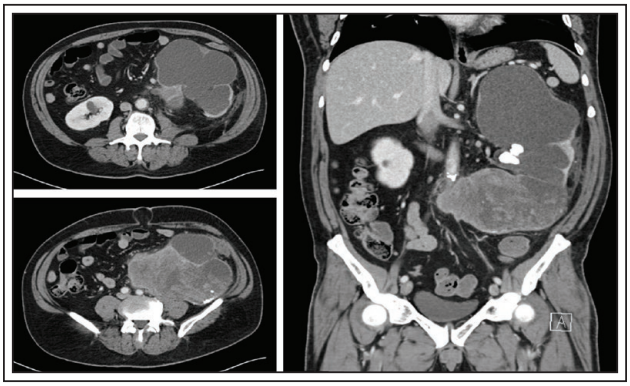

Figure 1. CT abdomen pelvis with contrast demonstrating left upper pole severe chronic hydronephrosis with large calculi and a heterogenous renal mass with dystrophic calcifications occupying the mid and lower poles with expansion outside of Gerota's fascia but not invading nearby organs suspicious for possible renal malignancy.

The initial plan was to manage the patient with an extended course of antibiotics with 3 weeks of sulfamethoxazole-trimethoprim per infectious disease (ID) recommendation, followed by left nephrectomy at a later date. However, he was re-admitted 2 weeks later due to worsening abdominal and back pain, loss of appetite, weight loss, and failure to thrive. During this admission a left percutaneous nephrostomy tube was placed for maximal drainage and 1100 mL of purulent brown urine was drained. Again his urine culture resulted no growth and the cytology was negative for malignant cells. He was discharged with an additional 2 weeks of amoxicillin-clavulanate per ID recommendations.

One month later, he underwent a robotic converted to open left radical nephrectomy. Conversion to an open approach was required due to significant adherence of the lower pole of the left kidney to the aorta and left common iliac artery as well as the left colon and small bowel. Additionally, general surgery was consulted intraoperatively for assistance with bowel mobilization and he ultimately required a left hemicolectomy due to dense adhesions of the colon and its mesentery to Gerota's fascia and the kidney. Pathology revealed a 12.5 cm mass arising from the renal pelvis invading into the renal parenchyma and peri-pelvic adipose tissue histologically consistent with moderately differentiated squamous cell carcinoma, Figure 2. The margins were negative, and three lymph nodes were negative for metastatic carcinoma. Final pathologic staging was pT3N0M0. Follow up CT of the chest/abdomen/pelvis at 2 months post-operation showed numerous new metastatic lesions within the liver, lung, spleen, anterior abdominal wall, and left nephrectomy bed. He was treated with palliative radiation to the abdominal wall and nephrectomy

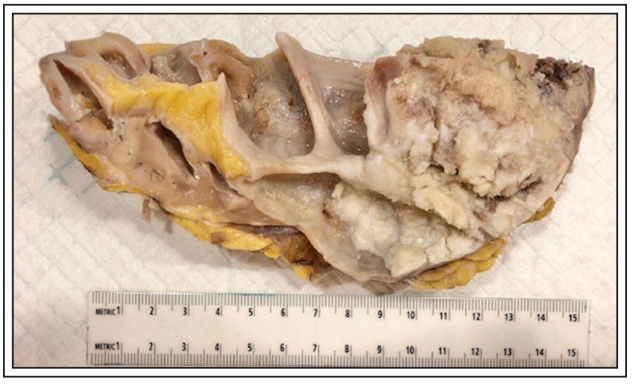

Figure 2. Gross pathology specimen: Left kidney showing squamous cell carcinoma (SCC) of the renal pelvis with invasion into the renal parenchyma.

bed due to pain and was planned to start palliative chemotherapy. However, he unfortunately rapidly deteriorated prior to initiation of chemotherapy, elected for transition to hospice care, and passed away shortly thereafter.

Discussion

This case study highlights the challenges of diagnosing a rare malignancy of the upper urinary tract. XGP and SCC of the upper tract urothelium can present with similar clinical signs and symptoms, including flank pain, nausea, vomiting, hematuria, weight loss, and malaise.[4-6] On radiographic imaging, XGP can often be mistaken for a variety of other pathologies including renal cell carcinoma, urothelial tumors, malakoplakia, and tuberculosis, hence earning the nickname "the great imitator".[5] Additionally, previous case reports have shown concurrent neoplasms with XGP pathologically after nephrectomy, further adding to the diagnostic confusion.[7] Given the rarity of upper tract SCC, it has a low pretest probability, and thus a radiographic diagnosis of XGP is typically higher on the differential.[1,2,5] Upper tract SCC and XGP are also both often associated with nephrolithiasis, with nephrolithiasis present in 14%-50% of patients with squamous cell carcinoma in the upper urinary tract, further confounding the picture.[8] The radiographic findings for SCC of the upper tract have been described in previous case reports as a solid renal pelvic or ureteric mass, hydronephrosis, calcifications or regional lymphadenopathy, which are vague findings that often overlap with XGP.[2,4,5] The vast majority of previously reported cases in the literature are based on CT as the modality of choice for cross sectional imaging. In our case, both CT and MRI were utilized, which did not yield any greater diagnostic accuracy.

Beyond radiographic studies, additional diagnostic modalities can be utilized in an attempt to differentiate these conditions, such as urine culture and cytology. However, this often does not clarify the clinical scenario. Although it may be assumed that a urine culture would be positive in XGP, studies show the positive culture rate in confirmed XGP cases to be only approximately 50%.[9] Voided urine cytology is a helpful adjunct in the detection of upper tract urothelial carcinoma, but data is limited on the clinical utility of urine cytology for upper tract SCC. It is possible to detect keratinization and keratotic cellular debris of SCC in preoperative urine cytology, but this remains insensitive diagnostic modality.[8] Although slightly more invasive, ultrasound guided fine needle aspiration cytology (FNAC) has been shown to aid in diagnosis of renal SCC.[10]

There is limited data on the treatment of SCC of the upper tract and cases in the literature have often highlighted a poor prognosis, as patients tend to present at an advanced stage.[5] There is currently no consensus on treatment, but most patients are treated with either nephrectomy or nephroureterectomy. There is currently limited data on the role of postoperative chemotherapy or radiation for metastatic lesions. Unfortunately, there is often significant renal damage and extrarenal metastasis at the time of diagnosis, making chemotherapy and radiation inefficacious after surgical resection.[3,5] Other case studies have utilized chemotherapy, but without any clear survival benefit.[5,6]

In conclusion, SCC of the renal pelvis is a rare but highly morbid malignancy that is difficult to diagnose, as it can be mistaken for XGP given its similar clinical presentation and radiographic findings. Our study is similar to previously reported case studies with SCC being discovered incidentally only after radical nephrectomy despite an extensive preoperative work up. Although uncommon, SCC should remain on the differential for patients with presumed XGP or those with longstanding renal calculi as it can affect patient counseling and surgical approach. □

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