Squamous Cell Carcinoma of the Renal Pelvis

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CASE PRESENTATION

The patient is a 46-year-old female with a history of chronic kidney disease and bilateral nephrolithiasis status post extracorporeal shockwave lithotripsy and multiple endoscopic procedures who presented with a malignancy of unknown origin. Approximately 1 month before her current admission, the patient underwent an elective right percutaneous nephrolithotomy. Three weeks after her discharge, the patient presented to an outside hospital with fever, chills, vomiting, and abdominal pain. She was admitted and treated for pyelonephritis. Cross-sectional imaging, completed during her hospital stay, was significant for hilar and mediastinal adenopathy, as well as a suspicious lung infiltrate. Fine needle aspiration of the mediastinal adenopathy was performed, and pathology was consistent with a poorly differentiated squamous cell carcinoma (SCC). Subsequently, the patient was referred to our institution for further evaluation and treatment.

On transfer, the patient reported persistent right flank pain because of her nephrostomy tube. She denied abdominal pain, fever, chills, nausea, vomiting, gross hematuria, or dysuria. Her medical history was significant for hypertension, nephrolithiasis, chronic renal insufficiency, and liver disease. Her surgical history was significant for multiple stone procedures, 1 caesarean section, and a tubal ligation. She denied any personal smoking history or history of living with a smoker. She denied any alcohol consumption or illicit drug use. Physical examination did not reveal any abdominal tenderness, distention, or palpable masses. She had mild right tenderness at the insertion site of the Malecot re-entry catheter. A complete blood count on admission revealed leukocytosis of 29,200/μL, thrombocytosis of 711 k/μL, and hemoglobin of 8 g/dL. Her serum chemistries demonstrated a sodium level of 134 mEq/L, a potassium level of 5.5 mEq/L, and a calcium level of 11.3 mg/dL, and her creatinine was at baseline of 2.4 mg/dL. Her alkaline phosphatase was 277 U/L, gamma glutamyl transferase was 330 U/L, and lactate dehydrogenase was 245 U/L.

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Noncontrast cross-sectional imaging of the abdomen and pelvis demonstrated marked enlargement of the right kidney (Figs. 1, 2). Dysmorphic, severely dilated renal collecting systems containing large, coarse calculi, renal cortical thinning, and perinephric stranding were present bilaterally. Curvilinear calcifications lined the dilated right lower pole collecting system. The ureters were not dilated. There was mild circumferential thickening of the urinary bladder wall. At the level of the renal hilum, several enlarged retroperitoneal lymph nodes were present.

Her hospital course was complicated by fevers of unknown origin and a persistent leukocytosis. Urine cultures were positive for *Enterobacter aerogenes*. Persistent, severe dilatation of the right lower pole collecting system prompted placement of an additional nephrostomy tube. Urine obtained directly from the nephrostomy tube was also sent for culture and cytology, but returned negative. There was some concern over possible tuberculosis, but despite antibiotic and empiric antituberculous therapy, the patient failed to improve, and additional sources of infection were investigated. Because of persistent tachycardia and worsening cardiac status, the patient underwent echocardiogram which revealed a small to moderate pericardial effusion. The patient underwent drainage of the effusion and a pericardial biopsy. No organisms were grown from the culture of the effusion, and the biopsy results reflected inflammatory changes.

DIFFERENTIAL DIAGNOSIS

Our differential diagnosis included xanthogranulomatous pyelonephritis (XGP), SCC, pyonephrosis, chronic pyelonephritis, and a renal cell carcinoma with paraneoplastic syndrome. Also, renal replacement lipomatosis was a consideration.

XGP is an atypical chronic bacterial pyelonephritis that can mimic renal cell carcinoma radiographically.[1] Microscopically, lipid-laden macrophages are the predominant cell in this reactive tissue lesion. The disease is not infrequently associated with diabetes mellitus and previous urologic surgery.[1] Women, typically middle-aged, are affected 3 times as frequently as men. Patients with XGP may have constitutional symptoms such as weight loss and anorexia.[1] A syndrome of liver dysfunction may occur in XGP and renal cell carcinoma. To further confuse the diagnosis, SCC is frequently associated with urolithiasis and hydronephrosis, and has been associated with paraneoplastic syndromes such as

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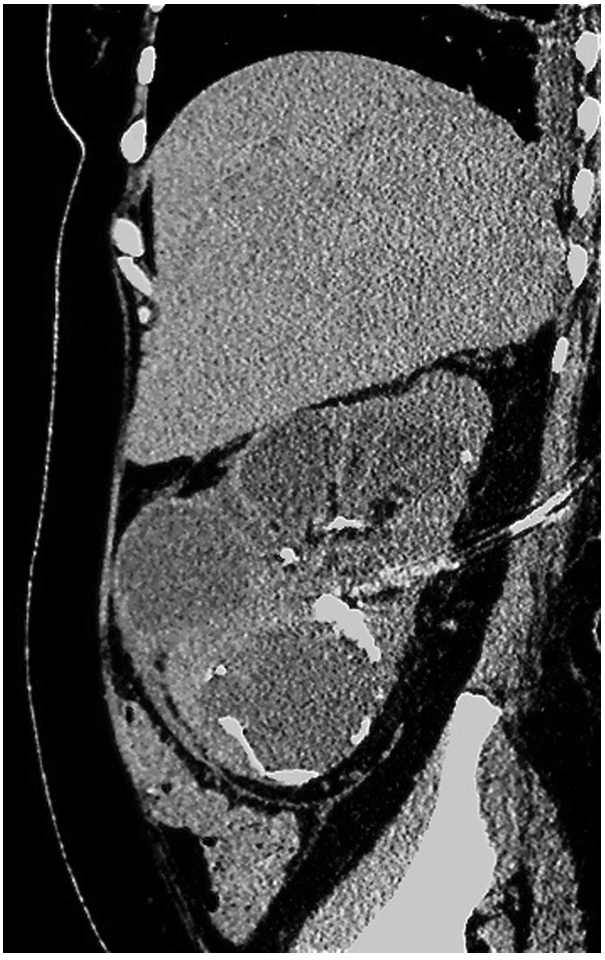

Figure 1. Saggital view of dysmorphic, severely dilated renal collecting systems containing large, coarse calculi, renal cortical thinning, and perinephric stranding.

hypercalcemia, leukocytosis, and thrombocytosis.[2] A solid mass, hydronephrosis, and calcifications are common radiological findings but nonspecific, which may explain why the diagnosis is not frequently established before the histopathologic examination of the surgical specimen.[2]

Chronic pyelonephritis is a condition that affects patients with chronic obstruction, secondary to nephrolithiasis or structural defects. As opposed to acute, nonobstructive pyelonephritis, chronic pyelonephritis has a propensity to lead to an end-stage renal disease.[3] Many times, there is absence of symptoms until renal insufficiency occurs. Histologically, there are patchy parenchyma changes, including parenchymal thinning and scar formation.[4]

Renal replacement lipomatosis and XGP have similar etiopathogenic, clinical, and radiological features. Both are characterized by atrophy and destruction of renal parenchyma, often associated with unilateral chronic renal infection, hydronephrosis, or pyonephrosis, and calculous disease.

MANAGEMENT, PATHOLOGIC REVIEW, AND OUTCOME

Because extensive laboratory and imaging evaluation failed to reveal another potential source for the patient's

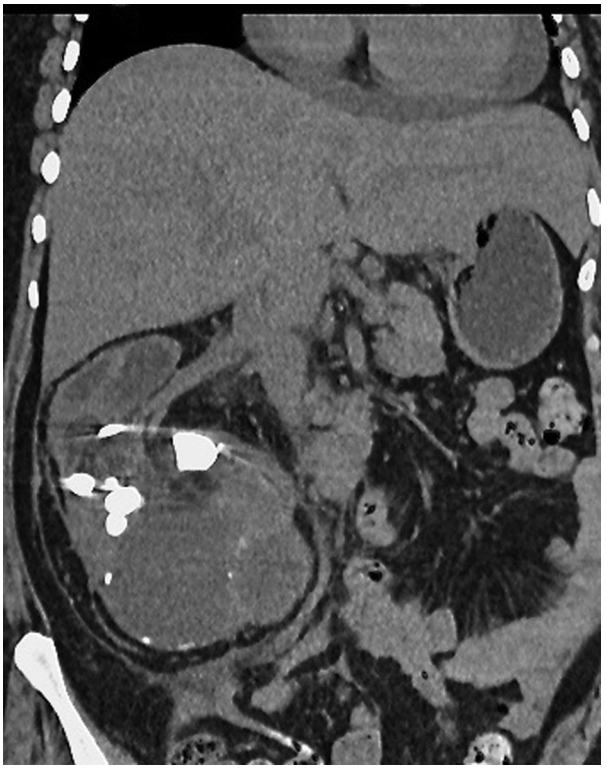

Figure 2. Coronal view of dysmorphic, severely dilated renal collecting systems containing large, coarse calculi, renal cortical thinning and perinephric stranding.

leukocytosis and fevers, the patient's right kidney became the presumed etiology for her clinical presentation. The patient failed to improve with conservative management, and ultimately she underwent a right open nephrectomy.

Intraoperatively, 1200 mL of ascitic fluid was aspirated on entry into the peritoneum. Extensive perirenal inflammation required significant time for mobilization and removal of the right kidney. The liver and the peritoneum were studded with metastatic implants. A sample was sent for frozen section analysis, consistent with poorly differentiated carcinoma.

The pathologist evaluated the right kidney (15.0 × 9.5 × 6.0 cm) with perinephric fat (800 g). Gross inspection revealed a 9.0 × 9.0 × 6.0 cm yellow-tan tumor involving the mid and lower pole of kidney parenchyma, calyces, renal pelvis, and adjacent perinephric adipose tissue without an extension to Gerota's fascia. Numerous black or green calculi (range, 0.2-1.5 cm) were present predominantly in the calyces (Fig. 3).

The histopathologic diagnosis was a moderately differentiated SCC with foci of glandular differentiation (Fig. 4) and foci of necrosis. The mucosa of the renal pelvis and calyces showed squamous metaplasia with areas of moderate and severe dysplasia. The tumor originated from the dysplastic squamous metaplastic epithelium lining the lower pole renal pelvis and/or calyces. Surgical margins were negative. The tumor was staged as pT4NxMx.

Postoperatively, the patient was extubated without incident. Her fevers eventually resolved, however, her