Case Report

Tsung-Hsin Chang, Jen-Shu Tseng*

Rare squamous cell carcinoma of the kidney with concurrent xanthogranulomatous pyelonephritis: A case report and review of the literature

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Abstract

Case presentation – In the current study, we report a 69-year-old female patient who was initially diagnosed with xanthogranulomatous pyelonephritis (XGPN) with nephrolithiasis and a peri-renal abscess. She presented to our department with right flank pain. Physical examination revealed right costovertebral angle knocking pain and computed tomography revealed dilated calyces and one staghorn stone over right kidney, with multiple abscess accumulations over the right peri-renal region. Right radical nephrectomy was performed using a transperitoneal flank approach, and pathology revealed squamous cell carcinoma (SCC) with concurrent XGPN. The patient was alive at 4 months post-operative follow-up. To the best of our knowledge, this is only the fifth case of renal SCC with concurrent XGPN reported in the English medical literature.

Conclusion – Renal SCC with coexisting XGPN is an extremely rare presentation and only four cases have been previously reported in the English medical literature. A positive diagnosis for this rare combination of diseases was established, based on pathological and immunohistochemical examinations after radical nephrectomy. Poor prognosis has been reported in such cases. Malignancies should be considered in patients with a long-standing history of urolithiasis.

Keywords: kidney, renal squamous cell carcinoma, xanthogranulomatous pyelonephritis

1 Introduction

Squamous cell carcinoma (SCC) of the kidney is extremely rare and accounts for less than 1% of all malignant renal tumors [1]. It is believed to be associated with the presence of urolithiasis and chronic infection [2–4]. Xanthogranulomatous pyelonephritis (XGPN) is also a rare disease resulting from chronic kidney infection secondary to obstructive uropathy, and both diseases share similar risk factors. However, the coexistence of SCC and XGPN is exceedingly rare and only a few cases have been previously reported. Treatment usually involves a radical nephrectomy and a positive diagnosis is made with the help of post-operative immunohistopathological examinations. A poor prognosis has been reported in previous studies [3]. The current study reports the clinical and pathological characteristics of renal SCC with concurrent XGPN. To the best of our knowledge, this is only the fifth report of renal SCC with concurrent XGPN in the English medical literature.

2 Case report

A 69-year-old female with a previous medical history of renal stones, recurrent urinary tract infections, diabetes mellitus (DM) and schizophrenia presented at our emergency department with right flank pain and right lower limb swelling for several days. She denied any alcohol, tobacco or substance abuse or addiction. There was also no associated family history including DM, urolithiasis or malignancies. Physical examination revealed right costovertebral knocking pain, local heat and tenderness over the right flank area with extension to the right thigh. Urine examination revealed 45 white blood cells per high-power field. Laboratory tests revealed leukocytosis with a total white blood cell count of 28.4 × 10³ μL (normal range: 4.0–10.0 × 10³ μL), elevated C-reactive protein level of 15.24 mg/dL (normal range: 0.0–0.79 mg/dL) and anemia with hemoglobin level of 128–133
8.7 g/dL (normal range: 11.0–16.0 g/dL). Renal and liver function test results were within normal ranges.

Computed tomography (CT) revealed a deformed right kidney with heterogeneously enhanced soft tissue and a staghorn stone occupying the renal pelvis, with hydronephrosis and mild hydroureter (Figure 1). Extensive abscesses were observed, which involved the right peri-renal space, psoas muscle, iliopsoas muscle and muscular layer of the abdominal wall, and extended down to the medial compartment of the quadriceps. After an initial survey, the patient was hospitalized. The initial diagnosis was XGPN with a possible connection to the multiple abscesses observed in the right half of the abdomen and thigh.

The patient received CT-guided drainage for the peri-renal and right thigh abscesses on the day of admission. Urine and drainage fluid culture both reported growth of Klebsiella pneumoniae and Escherichia coli; the intravenous antibiotics such as Brosym and Tigecycline were chosen according to the patient’s sensitivity test results. Right nephrectomy was performed via a transperitoneal flank approach after ten days of hospitalization. The patient was placed in the left lateral position with a raised kidney bar and a 15 cm right lumbar incision was made inferior to the 12th rib. About 300 mL of pus fluid was found and drained from the peri-renal space. The original surgical plan was a retroperitoneal nephrectomy. However, due to the presence of severe adhesions, the peritoneal space was exposed. Careful dissection was made along the Gerota’s facia, and dissection of the hilar region and kidney margins was performed with a 45 × 2.5 Endo-GIA™. The ureter was then transected with an Endo-GIA™ and the whole kidney was removed. A no. 19 channel drain was placed in the renal fossa.

The resected specimen weighed 333 g and measured 11.2 × 7.6 × 6.6 cm in size (Figure 2). The kidney capsule was thickened and adhered to the cortex and peri-renal fat, while the subcapsular surface was smooth. When the specimen was sectioned, it was observed that the pelvis and calyces were markedly dilated with thinning of the renal parenchyma. The cortex measured up to 0.2 cm in thickness. The calyces and pelvis were distended due to obstruction of the renal pelvis by a staghorn stone.

Microscopic examination revealed a moderately differentiated SCC with extensive keratin deposition and pearl formation (Figure 3). No normal renal cortical tissue was identified. The adjacent parenchyma showed focal areas of lipid-laden macrophage (foamy histiocytes) distribution intermixed with lymphocytes, giant cells and fibrocollagenous tissue. Immunohistochemically, the tumor cells were positive for p63 and negative for GATA-3 (Figure 4). Thus, based on the morphology and immunohistochemical studies, a diagnosis of moderately differentiated renal SCC with focal XGPN

Figure 1: Patient CT. CT imaging revealed. (a) A staghorn stone (red arrow) occupying the renal pelvis with dilated calyces and hydronephrosis and (b) heterogeneously enhanced soft tissue (red arrow) with extensive abscesses (*) over the right abdomen.
associated with urolithiasis was made. The infection improved after the surgery. The patient did not receive any additional adjuvant treatment for SCC according to the patient’s and her family’s will. The patient is still alive and performing daily activities at 4 months post-operative follow-up. There were no recurrent masses or metastatic lesions found in the follow-up echogram or CT imaging.

Informed consent was obtained in both written and verbal format from the patient and her family to publish this case report and any accompanying images.

3 Discussion

Renal SCC is a rare malignancy with a reported incidence rate of 0.5–8% [1,3]. Generally, these tumors are expected to have poor clinical outcomes due to their aggressiveness and typically high disease stage at diagnosis [3]. The majority of tumors are locally advanced or metastatic when detected, and thus at a histologically high grade [5]. Hematuria, pain and local symptoms are common and this lack of symptom specificity may cause a delay in diagnosis [3]. The
mean delay from initial presentation to surgery was 51.5 days according to previous data [3]. Risk factors for renal SCC have been reported to be chronic pyelonephritis [1–3], chronic renal calculi irritation [6], phenacetin consumption [7] and squamous metaplasia in response to chronic irritation [3,6]. Solid mass, hydronephrosis and calcification are common but nonspecific findings observed in radiological examinations.

Making the correct tentative diagnosis from limited and highly imitative clinical images can be difficult. A positive diagnosis of renal SCC is difficult and primarily relies on immunohistochemical examinations following surgery. The main differential diagnoses for renal SCC are urothelial carcinomas (UCs), other renal neoplasms and XGPN. Establishing a correct diagnosis of SCC can be challenging since UC may also present with focal squamous differentiation. Such cases are best classified as UC with squamous differentiation. Thorough examination by pathology specialists is required to ensure that whole histological specimens contain only pure SCC without a combination of UC or another renal neoplasms, in order to make the correct diagnosis.

XGPN is also a rare, severe, chronic renal infection and it is often associated with diffuse renal destruction. It has been reported that the primary pathogeneses associated with XGPN are nephrolithiasis, obstruction and infection [8]. It has been known as “the great imitator” [9] due to its possible resemblance to every inflammatory disease of the kidney, as well as renal neoplasms, on radiographic examinations and in its clinical manifestations [9].

Keratinizing squamous metaplasias may develop as a result of a long-standing history of calculi. There have been previous reports on XGPN with concurrent squamous metaplasia [10], but only a few reports where metaplasias actually lead to SCC. The coexistence of renal SCC and XGPN is extremely rare in the previously published literature, and there are only a few reports of such lesions on record (Table 1). To the best of our knowledge, this is only the fifth case to be reported in the English medical literature.

The current consensus on the best primary treatment for such cases is a radical nephrectomy with close post-operative follow-up. A chemotherapy regimen of methotrexate, cisplatin and bleomycin may have a little but short effect when the disease becomes metastatic [11]. Makino et al. [12] reported on neoadjuvant chemotherapy for SCC of the upper and lower urinary tracts but their results showed no significant differences in cancer-specific survival and the benefits remain uncertain due to the limited and poor efficacy.

4 Conclusion

In conclusion, renal SCC and XGPN are both rare diseases associated with chronic kidney infection and long-standing obstructive uropathy. Renal SCCs with coexisting XGPN are very rare and only four cases have been previously reported. Surgical treatment is often preferred and a definite diagnosis is made with the help
of immunohistochemistry post-surgery. Perioperative chemotherapy showed poor efficacy and uncertain survival benefits. Poor outcomes have been reported in previous cases. We hereby report this case to highlight that malignancies should be considered in patients with an extensive history of infected staghorn calculi, and this may also be a reason for urologists to operate on asymptomatic staghorn stones before it is possible for them to develop into neoplasms.

**Conflict of interest:** The authors report no conflicts of interest.

**Data availability statement:** The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

**References**


