Incidently detected squamous cell carcinoma of the renal pelvis in patients with staghorn calculi — A case report and literature review

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Article history:
Received 18 September 2012
Received in revised form 13 November 2012
Accepted 25 February 2013
Available online 18 June 2013

Abstract
Squamous cell carcinomas of the renal pelvis (renal squamous cell carcinoma, RSCC) are rare. They are highly malignant tumors of urothelial origin and have a poor prognosis owing to misleading clinical presentations and difficulty in early diagnosis. We present a case of incidentally detected RSCC in patients with staghorn calculi and discuss this entity.

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1. Introduction
Cancers of the renal pelvis are uncommon. Most of them are urothelial carcinomas, and about 10% are squamous cell carcinomas.1 The etiological factors of RSCC are believed to be chronic inflammation and infection with longstanding urolithiasis.2 The diagnosis of RSCC is usually challenging owing to its insidious onset, misleading clinical manifestations, and nonspecific imaging survey. We present a case of incidentally detected RSCC in a patient with staghorn calculi and discuss this entity.

2. Case report
A 69-year-old male who had a past history of bilateral renal stones for many years and chronic kidney disease (Grade III) was admitted to our hospital several times because of recurrent urinary tract infection. Right percutaneous nephrolithotomy had been performed on December 13, 2007. (Fig. 1A and B). On this occasion, he presented with poor appetite and general weakness for 1 week. He also complained of mild fever, bilateral flank pain, dysuria, frequency, nausea, and body weight loss. Physical examination revealed bilateral flank knocking pain, and ultrasonography showed bilateral renal stones with hydronephrosis. Leukocytosis, elevated C-reactive protein (CRP), pyuria, and microscopic hematuria were also noted. Bilateral acute pyelonephritis was impressed, and antibiotics with ciprofloxacin were administered on the basis of previous urine culture (Stenotrophomonas maltophilia). However, spiking fever and hypotension occurred, and we changed the antibiotics to ertapenem. Bilateral percutaneous nephrostomies were also inserted to release hydronephrosis, and pus-like fluids were drained away from the right kidney. The urine cytology collected from bilateral percutaneous nephrostomies showed a negative result. Nonenhanced computed tomography (CT) scan of the abdomen was carried out and revealed bilateral renal stones with the right kidney appearing edematous and enlarged with surrounding perinephric fat stranding. High-density nodules occupying the right renal calices (CT number: 35HU) were also noted,
and differential diagnoses included hematoma, renal tumor, and xanthogranulomatous pyelonephritis (XGPN; Fig. 1C). After consultation, right nephrectomy was performed on April 22, 2010, and the specimen showed multiple renal stones occupying the renal calyces with pus-like fluid accumulation and a bulky, white, central nodular tumor with renal parenchyma invasion was also noted (Fig. 2). The pathology revealed the kidney parenchyma was diffusely infiltrated by keratinizing squamoid neoplastic cells and...
keratin pearls were also noted; consequently, RSCC was impressed with the pathology stage: T3NxMx (Fig. 3). After the operation, fever and flank pain subsided, and the patient was discharged after 2 weeks of treatment. However, 5 months later abdominal CT showed multiple liver metastases and systemic chemotherapy with carboplatin and fluorouracil (5-FU) was administered twice. He expired 6 months after the operation.

3. Discussion

It is difficult for the clinical physician to distinguish RSCC from granulomatous reaction to chronic infection, such as XGPN3-5 and it is believed that chronic irritation, inflammation, and infection induce squamous metaplasia, which may subsequently develop into squamous cell carcinoma. They share similar clinical symptoms, consisting of fever, flank pain, hematuria, obstructive uropathy, body weight loss, low urinary tract symptoms, and similar etiological factors. Most cases occur in the setting of massive destruction of the kidney, the appearance may be misleading, thus leading to confusion in proper diagnosis.

To date, there is still no standard examination to differentiate these two diseases except via pathological scrutiny. However, experts have attempted to determine the key in differentiation and have placed emphasis on imaging studies. In 1983, Wimbish et al6 described a case report about the sonography and CT appearance of RSCC. The sonography revealed a solid, slightly hyperechoic material in the calyx, and CT showed a mass of slightly higher attenuation than renal parenchyma, infiltrating between the low-attenuation areas and within the renal pelvis. The presence of calculi may also be helpful in suggesting the diagnosis. The magnetic resonance imaging (MRI) also has benefits for differentiations. In MRI, the RSCC may show as a solid mass, hydronephrosis, or calcifications, and XGPN appears high-intensity signal on T1-weighted images due to the accumulation of lipid-laden foamy macrophages. However, angiomyolipoma, retroperitoneal liposarcoma, renal cell carcinoma occasionally contain fatty tissue and are therefore extremely difficult to differentiate from XGPN.7

In 2003, Lee et al8 published a retrospective review of 26 cases with RSCC and two radiographic signs were presumed as follows: the “separating nephroliths sign” and the “distorted nephroliths sign.” The alteration of distance or arrangement between renal stones in two sequential radiographs was defined as the “separating nephroliths sign,” and the irregular border of renal stones in a cluster pattern distorted in a single radiograph was considered to be the “distorted nephroliths sign.” The incidences of “separating nephroliths sign” and “distorted nephroliths sign” were 72% (8 positives in 11 cases) and 20% (3 positives in 15 cases), respectively. The overall positive rate was 39% among all cases of RSCC.8

The diagnosis of RSCC is confirmed by pathology from nephrectomy or percutaneous renal biopsy. Grossly, it is usually large,
necrotic, and ulcerated, with gross invasion of the renal parenchyma and retroperitoneal soft tissues. Histologically, it shows atypical keratinizing or nonkeratinizing squamous neoplastic cells, but the characteristics of squamous cell carcinoma (such as keratin pearls and intercellular bridges) may not be apparent because most of the cases of RSCC are moderately or poorly differentiated. There is no evidence of diagnostic values in urine cytology and serum SCC level in the literatures.9,10

The treatments of renal pelvis squamous cell carcinoma include radical nephrectomy/nephroureterectomy, radiotherapy, and neo-adjuvant/adjuvant systemic chemotherapy. However, on initial diagnosis, it is high stage in most cases (pt3 or higher in >95% of cases), which is associated with a poor prognosis. The median survival is 7 months after surgery and a 5-year survival rate of less than 10.1 Nephrectomy/nephroureterectomy may sometimes result in cure in low-stage patients, but is rarely curative in the high stage. Adjuvant radiation and chemotherapy are generally ineffective. There are few reports of RSCC of the renal pelvis in the medical literature (Table 1),9 and new treatment modalities are urgently needed to improve the poor prognosis in patients with advanced-stage squamous cell carcinoma.

Tracing back to our patient, he had a past history of bilateral urolithiasis and recurrent urinary tract infection. He had regular follow-up in our clinical department, but we were unable to find the “separating nephroliths sign” and the “distorted nephroliths sign” in the radiography owing to previous surgical intervention distorting the arrangement of renal stones. By contrast, the CT scan told us that there were high-density nodules with slightly higher attenuation than renal parenchyma occupying the renal calices (CT number: 35HU); therefore, differential diagnoses included XGPN, hematoma, and renal tumor. Percutaneous renal biopsy was not considered in our case owing to the risk of seeding and the high false-negative results.3 We performed the nephrectomy for the sake of infection control, but eventually, RSCC was found. The difficulty in differentiation between RSCC from XGPN preoperatively is noted in our case.

It is often not possible to distinguish RSCC from XGPN preoperatively. In clinical practice, false diagnoses are therefore frequent. We herein present a case of RSCC mimicking XGPN, and recommend that sonography or CT should be performed without delay to detect any possibility of malignancy.

Conflicts of interest statement

The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in the manuscript.

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