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Case Report

Renal squamous cell carcinoma mimicking xanthogranulomatous pyelonephritis: Case report and review of literature

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ARTICLE INFO

Article history: Received 18 January 2016 Accepted 28 February 2016 Available online 4 April 2016

Keywords: Renal squamous cell carcinoma Xanthogranulomatous pyelonephritis Kidney

ABSTRACT

Primary renal squamous cell carcinoma (SCC) is a rare primary malignancy of the kidney. Diagnosis is usually delayed because of its lack of characteristic clinical and imaging features and inherent aggressive nature. We present a case of primary renal SCC in a 66-year-old woman with bilateral renal calculi and a complex right lower pole renal mass. The diagnosis of primary renal SCC was established based on the histopathology after right nephrectomy. Copyright © 2016, the Authors. Published by Elsevier Inc. under copyright license from the University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Introduction

Primary renal squamous cell carcinoma (SCC) is an extremely rare entity, and it comprises less than 1% of all urinary tract neoplasms [1]. Because of its lack of characteristic presentation, such as palpable mass, hematuria, and pain, the patients usually present late resulting in delay in diagnosis [2]. Urolithiasis and hydronephrosis are often associated with renal SCC [3]. It is postulated that chronic irritation of the renal pelvis results in squamous metaplasia, which later increases the risk of developing into SCC [4]. Renal SCC has poor prognosis with few surviving more than 5 years due to its early metastatic spread [3]. We present a rare case of a primary renal SCC in a 66-year-old woman.

Case report

A 66-year-old gentleman with history of hypertension presented to our institution with bilateral flank pain and hematuria for 3 days associated with oliguria. There was also associated weight loss and anorexia. Physical examination was unremarkable. Laboratory tests revealed elevated serum creatinine of 780 umol/L (normal range: 60-105 umol/L) and urea of 33.4 mmol/L (2.9-9.3 mmol/L), hyponatremia with a serum sodium of 119 mmol/L (135-144 mmol/L), anemia with hemoglobin level of 8.9 g/dL (13.0-17.0 g/dL), and leukocytosis with total white cell count of 13.2×10^9 /L (3.6-9.3 x 10^9 /L). C-reactive protein was raised at 64.5 mg/L (0.0-5.0 mg/L).

Written informed consent for the case to be published (including images, case history, and clinical information) was obtained from the next-of-kin for publication of this case report.

Competing Interests: The authors declare that there are no competing interests regarding publication of this article.

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Plain abdominal radiograph showed a right staghorn calculus and several smaller right renal stones (Fig. 1). Unenhanced computed tomography (CT) of the kidneys, ureters, and bladder confirmed a staghorn calculus with right hydronephrosis. There was also an irregular infiltrative right lower pole renal mass invading the right psoas muscle (Fig. 2).

Contrast-enhanced magnetic resonance imaging (MRI) of the kidneys was performed to delineate the right renal mass. T2-weighted imaging showed an irregular mass of low-to-intermediate signal centered in the parenchyma of the lower pole of the right kidney invading the right psoas muscle (Figs. 3 and 4). T1-weighted imaging showed the mass to by slightly hyperintense (Fig. 5). The mass demonstrated restricted diffusion (Fig. 6). Postcontrast, there was peripheral enhancement with nonenhancing central areas (Fig. 7).

Initial impression was xanthogranulomatous pyelone-phritis (XGP). However, an infiltrative malignancy could not be excluded on imaging and ultrasound-guided biopsy of the lesion was performed. Histologic examination showed small fragments of necrotic tissue and squamous epithelium with parakeratosis and hyperkeratosis. In view of a lack of definite histology from biopsy, the patient underwent an open right radical nephrectomy. Histologic examination of the nephrectomy specimen revealed features of keratinizing SCC associated with squamous metaplasia and squamous carcinoma in situ. Final diagnosis was confirmed to be SCC of the kidney.



Fig. 1 – Plain radiograph of the abdomen showed multiple right renal stones.



Fig. 2 — Coronal CT showed, in addition to right renal stones, an infiltrative mass in the lower pole of right kidney invading the right psoas (arrow). Further characterization was not possible in the absence of intravenous contrast.

The patient had a stormy postoperative period complicated by sepsis, end-stage renal failure requiring dialysis, and new liver metastasis. In view of poor prognosis, he was terminally discharge with best supportive care. He eventually passed away at home a month later.

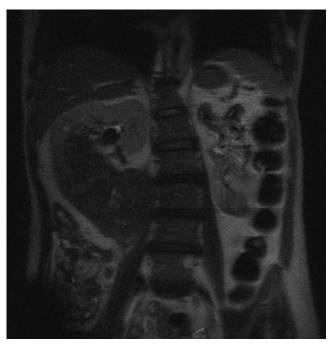


Fig. 3 — Coronal T2-weighted sequence showed a predominantly T2-hypointense, irregular mass in the lower pole of the right kidney invading the right psoas muscle.

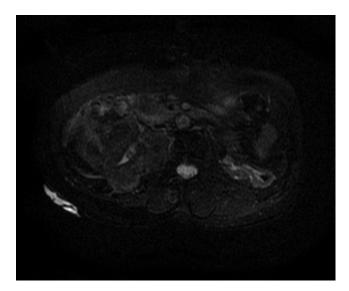


Fig. 4 — Axial T2-weighted fat-saturated sequence again showed the irregular mass in the lower pole of the right kidney to be T2-hypointense. Invasion of the right psoas muscle is again demonstrated.

Discussion

Renal SCC is a rare renal malignancy. It is usually centered in the renal pelvis and secondarily invades the renal sinus fat and renal parenchyma [2]. Renal SCC is aggressive and high grade and usually advanced at the time of presentation, resulting in poor prognosis [5].

Urolithiasis, hydronephrosis, and paraneoplastic syndromes are often associated with renal SCC [3]. The etiologic factors of renal pelvis SCC are recurrent urinary tract infections with or without vesicoureteric reflux, long-standing

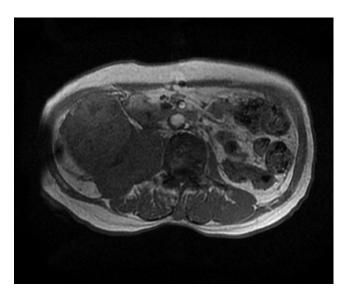


Fig. 5 — Axial T1-weighted sequence showed the irregular mass in the lower pole of the right kidney to be slightly T1-hyperintense.

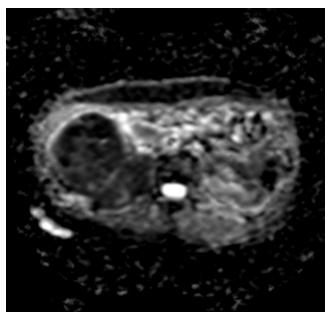


Fig. 6 – Apparent diffusion coefficient map showed the mass in the lower pole of the right kidney to be hypointense compatible with restricted diffusion.

staghorn calculi, smoking, schistosomiasis, exogenous and endogenous chemicals, vitamin A deficiency, and hormonal imbalance [4]. Although not statistically significant, Holmang et al. [3] found that history of abuse of analgesics containing phenacetin or previous surgery for urolithiasis more common among patients with SCC than urothelial carcinoma.

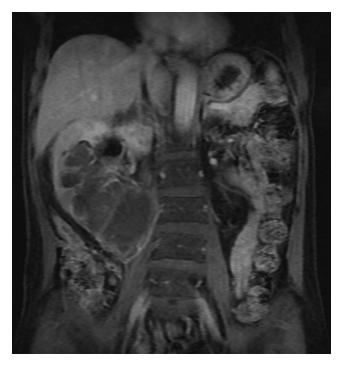


Fig. 7 — Postcontrast T1-weighted fat-saturated sequence showed irregular peripheral enhancement of the mass with nonenhancing areas centrally.

Although all cases of renal SCC are associated with renal stones, there are no specific-imaging features. Renal SCC can present as diffuse enlarged nonfunctional kidney with renal calculi, hydronephrosis, perirenal infiltration, and low density or echogenicity in the renal parenchyma [3]. It can present as infiltrative soft tissue in the renal pelvis without evidence of a distinct mass [1]. Solid-cystic masses have been described [6]. More specific findings on imaging would be enhancing extraluminal and exophytic mass [7]. These varying imaging appearances often contribute to delay in diagnosis until the availability of histopathologic examination of the surgical specimen.

The main differential diagnosis of renal SCC is XGP, an entity also associated with renal calculi. XGP is an uncommon form of chronic pyelonephritis resulting from chronic obstruction usually from renal calculi, which leads to hydronephrosis, formation of an inflammatory mass causing destruction of renal parenchyma, mimicking malignancy [2]. A typical feature of XGP on contrast-enhanced CT is the "bear-paw sign" is seen as low-attenuating areas surrounded by enhancing thinned-out renal parenchyma from chronic hydronephrosis [8]. However, this may not always be seen. Moreover, XGP can invade adjacent structures, which makes it even more difficult to distinguish from an aggressive malignancy [9].

In our patient, the presence of multiple renal stones, hydronephrosis, and an infiltrative renal mass suggested either renal XGP or an unusual aggressive malignancy. MRI showed a relatively T2-hypointense, mildly T1-hyperintense mass with peripheral enhancement, and nonenhancing central regions, which are features of XGP [10]. However, percutaneous biopsy obtained necrotic tissue and squamous epithelium, the latter being unusual in renal tissue. Renal SCC was only confirmed on nephrectomy. This emphasizes the need for high index of suspicion to differentiate renal SCC from XGP despite initial biopsy results.

Given its insidious onset of clinical symptoms with poor prognosis and strong association between renal SCC with urolithiasis, it is recommended that patients with known urolithiasis to be screened with imaging modalities to achieve early diagnosis. Lee et al. [7] recommends intravenous urogram to be performed periodically in patients with renal stones, particularly in those with long-standing history. Filling defect in the collecting system, delayed appearance of pyelogram, or renal parenchymal thickening should be regarded as a sign of renal tumor despite the absence of mass effect and preservation of smooth renal contour. This should prompt further evaluation with cross-sectional imaging such as CT or MRI.

Patients with SCC stage T1-T2 may be treated with radical surgery and have good prognosis although in at least 50% of patients, symptomatic residual tumor, or local recurrence seem to develop [3]. Adjuvant chemotherapy and radiotherapy can be given in advanced renal SCC but have not shown survival benefits [11]. The 5-year survival rate for

advanced renal SCC is less than 10% with the median survival time of 5 months [12].

Conclusions

This case report serves to highlight that the diagnosis of malignancy, in particular renal SCC should be considered in a patient having long-standing history of urolithiasis presenting with renal mass. XGP is a differential diagnosis. High index of suspicion on various imaging modalities and biopsy is important in distinguishing the 2 entities.

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