Case Report
High-fever as an initial manifestation of sarcomatoid carcinoma of renal pelvis: a case report and literature review

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Abstract: Background: Renal pelvis sarcomatoid carcinoma (RPSC) is a rare neoplasm originating from the urinary tract. The behavior of this tumor is often invasive, with a poor prognosis. Case presentation: A 63-year-old man presented with hyperpyrexia and left loin intermittent pain for a period of one month. Abdominal computed tomography revealed a 5.6 × 4.6 cm irregular mass with slight contrast enhancement, arising from the upper pole of his left kidney. He received a radical left nephrectomy. Pathologic examination disclosed sarcomatoid urothelial carcinoma, an unusual subtype of urothelial carcinoma with an invasive course. His postoperative course was uneventful and he was discharged on the eighth day. Conclusion: Hyperpyrexia and loin pain as the initial symptoms of RPSC is rare. This is the first case in which reported high-fever as a sign of sarcomatoid carcinoma of the renal pelvis in the literature.

Keywords: Renal pelvis sarcomatoid carcinoma, hyperpyrexia, paraneoplastic syndrome

Introduction

Sarcomatoid carcinoma is a rare neoplasm originating from both epithelial and mesenchymal tissues. It constitutes a variant of carcinoma, whose sarcomatoid component arises from the metaplasia of carcinoma tissue. Histologically, sarcomatoid carcinoma consists of two components: the epithelial and the sarcomatoid mesenchymal component. The tumor interstitium often consists of chaotic and randomly arranged fascicular spindle cells; however, as spindle cells can also be present in healthy tissues, the proportion of sarcomatoid tissues must exceed a pre-defined threshold so that a pathologic diagnosis of sarcomatoid carcinoma can be satisfied. Past reports have suggested that the percentage of sarcomatoid tissues must be higher than 50% in such scenario [1]. If the percentage is lower than 50%, the pathological diagnosis should be defined as carcinoma.

Sarcomatoid carcinoma has been reported to develop in organs including the kidney, the urinary bladder, and the female genital tract. Renal pelvis sarcomatoid carcinoma is an extremely rare clinical finding. We presented one such case in the following report.

Case presentation

A 63-year-old man presented with a one-month history of hyperpyrexia and abdominal pain without gross hematuria. Physical examination showed no significant abnormalities. Abdominal computed tomography (CT) ordered for evaluating his abdominal pain origin revealed a 5.6 × 4.6 cm neoplasm with slight contrast enhancement over his left renal pelvis (Figure 1). Contrast-enhanced abdominal ultrasound also demonstrated a 5.7 × 4.5 cm mass in the upper part of the left kidney with low enhancement, atypical for renal cell carcinoma (RCC). His intermittent hyperpyrexia was accompanied by a white blood count of 9.41 × 10⁹/L (normal range, 3.5-9.5 × 10⁹/L). He had no symptoms and signs of urinary tract infection. Findings from his chest CT suggested that his fever did not result from respiratory system abnormalities. In addition, anti-pyretic medication, including acetaminophen, could ameliorate his fever,
RPSC is a rare neoplasm originating from epithelial and mesenchymal tissues but antibiotic treatment was not efficacious. A biopsy of the mass revealed renal cell carcinoma, and the preoperative diagnosis was left RCC, cT1bM0N0. Other blood test results were normal, so antibiotic treatment was not initiated. We suspected that his high fever was related to the left renal tumor. He then received a left laparoscopic nephrectomy.

The resected tumor weighed 334 g with a volume of $11 \times 7.5 \times 7$ cm. The pathology report revealed a sarcomatoid urothelial carcinoma invading into renal sinus, parenchyma, perinephric fat, neural, and lymphovascular tissues

**Figure 1.** Contrast-enhanced computed tomography revealed a $5.6 \times 4.6$ cm mass with slight enhancement in left kidney.

**Figure 2.** The sarcomatous area is mainly composed of spindle cells (original magnification $\times 40$; hematoxylin & eosin staining).

(Figure 2). Immunohistochemical examination revealed positivity to CD10 (Figure 3), CK7 (Figure 4), CK-pan (+) (Figure 5), p63 (Figure 6), and vimentin (Figure 7).

The pyrexia subsequently resolved promptly after the primary tumor was removed. The patient refused further surgery aiming to remove a ureteral tumor remnant. We followed up the patient periodically. The patient relapsed during subsequent examinations, with multiple organ metastasis 6 months after the operation. He passed away 10 months after the operation.

**Discussion**

Through a comprehensive literature search using PUBMED, 24 similar cases were found [2-14]. The initial presentations of these 23 patients consisted of hematuria and loin pain
RPSC is a rare neoplasm originating from epithelial and mesenchymal tissues. It has been reported that malignant tumors may present with unexplained fever, and tumor fever is usually attributed to paraneoplastic syndrome (PNS) [15]. PNS results from tumoral secretion of functional peptides and hormones, or from immunologic cross-reactivity between tumor tissues and self-peptides. Many patients with RCC report PNS. Approximately 25% of patients with RCC present with fever, and fever can be the sole presentation in approximately 2% of patients. In our case, we found that his fever may be preferably treated as hyperthermia, and high fever can be the first symptom of RPSC. This fever played an important role in formulating our preliminary diagnosis.

Ruling out the possibility of tumor fever as early as possible is often required for its effective treatment. For patients, their family members and physicians, persistent fever can be associated with stress, waiting for effective treatment and excessive use of healthcare resources. Once the diagnosis of tumor fever is made, both surgical and medical oncological treatments can assist in controlling the fever. Indeed, naproxen and other non-steroidal anti-inflammatory drugs can effectively ameliorate fever. In our case, the pyrexia resolved shortly after tumor resection.

The prognosis for patients with a diagnosis of RPSC is poor. Most patients die within 6 to 12 months after surgery. Both domestic reports and those worldwide suggest that surgery is the most effective treatment modality. The type of operation of choice generally varies according to patient tolerability, including simple nephrectomy, or nephroureterectomy with or without bladder cuff excision.

In conclusion, we found that surgical resection should be provided to patients who can tolerate nephroureterectomy. The existing literature identifies that RPSC is insensitive to radiotherapy and chemotherapy, and early diagnosis followed by surgery is important to improve patient prognosis.

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Figure 5. Immunohistochemical staining revealed positivity to CK-pan (original magnification × 20).

Figure 6. Immunohistochemical staining revealed positivity to p63 (original magnification × 20).

Figure 7. Immunohistochemical staining revealed positivity to vimentin (original magnification × 20).
RPSC is a rare neoplasm originating from epithelial and mesenchymal tissues

the written consent is available for review by the editor of this journal.

Disclosure of conflict of interest

None.

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References


