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Malignancy of a Horseshoe Kidney: A Case Series with a Rare Presentation

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ABSTRACT

Horseshoe kidney is the most common fusion anomaly. Patients with horseshoe kidney typically present with symptoms related to infection, stone formation, and hydronephrosis. Rarely, patients may present with malignancy of horseshoe kidney, and most of them arise from isthmus. We are presenting a case series of 2 cases of renal cell carcinoma arising from horseshoe kidney.

INTRODUCTION

Horseshoe kidney (HSK) is the most common fusion anomaly, with an incidence rate of 1 in 400 of the general population [1]. While most of the reported cases are incidental findings, sometimes a patient may present with pictures of hydronephrosis, infection, and calculus formation [2]. Rarely, a patient presents with malignancy of the renal unit. Hildebrand, in 1895, is credited with reporting the first case of malignancy of HSK [3]. Most of these malignancies were arising from isthmus of HSK [4], and the majority of these were of the clear-cell variety. We are reporting here 2 cases of renal cell carcinoma (RCC), including a very rare case of the papillary-cell variety arising from the upper pole.

CASE REPORT

Case Number 1

A 50-year-old female patient presented to our outpatient department with a chief complaint of a single episode of painless, total gross hematuria not associated with the passage of clots 2 months before presentation. There was no history of flank pain, dysuria, fever, graveluria, weight loss, or anorexia. On physical examination, an ill-defined lump 7 cm x 5 cm was palpable in the upper right quadrant involving the right lumbar and umbilical regions. The baseline blood investigations were within the normal limit. His urine examination revealed 30-35 RBC/HPF. An ultrasonography of the abdomen indicated the presence of HSK with a mass approximately 7 cm x 6 cm arising from the upper pole of the right renal moiety.

The multidetecting computer tomography (CT) scan confirmed the presence of an HSK with fusion at the lower pole, an enhanced lesion of 7.3 cm x 7 cm arising from the upper pole of the right renal moiety, and areas of calcification and necrosis suggestive of RCC (Figure 1). The metastatic work-up, which included a chest X-ray and liver function test, was normal. On exploration, HSK with a well-formed isthmus connecting the lower pole of both kidneys with a mass arising from the upper pole of the right renal moiety was seen. No significant lymphadenopathy was noticed (Figure 2). The patient underwent right radical nephrectomy with isthmectomy by the right subcostal transperitoneal approach. Intraoperatively, the finding of a HSK with a mass arising from the right upper pole was confirmed. A well-formed isthmus was found connecting to the lower pole. The stump of the isthmus was run under with an absorbable running suture, and then it was covered with an omental patch to prevent hemorrhage and urinary fistula

KEYWORDS: Clear-cell carcinoma, fusion anomaly, horseshoe kidney, papillary-cell carcinoma, renal cell carcinoma

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ABBREVIATIONS AND ACRONYMS

HSK: Horseshoe kidney
LFT: Liver function test
OPD: Outpatient department
RCC: Renal cell carcinoma

Figure 1. A CT scan showing a mass arising from the right renal moiety of a horseshoe kidney. .

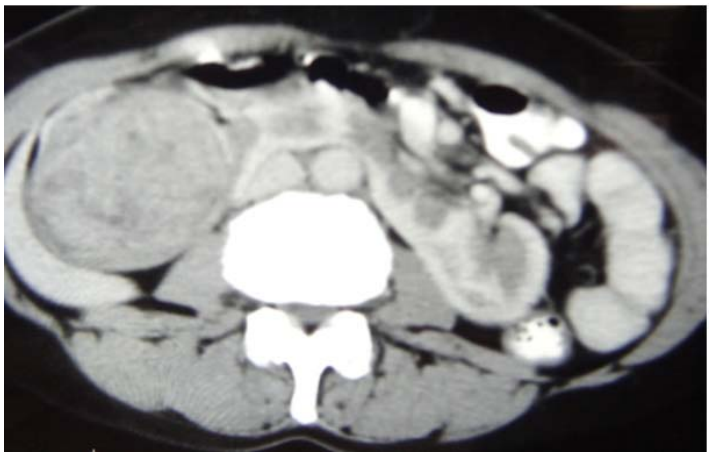


Figure 2. An intraoperative picture of the right renal moiety with the isthmus.

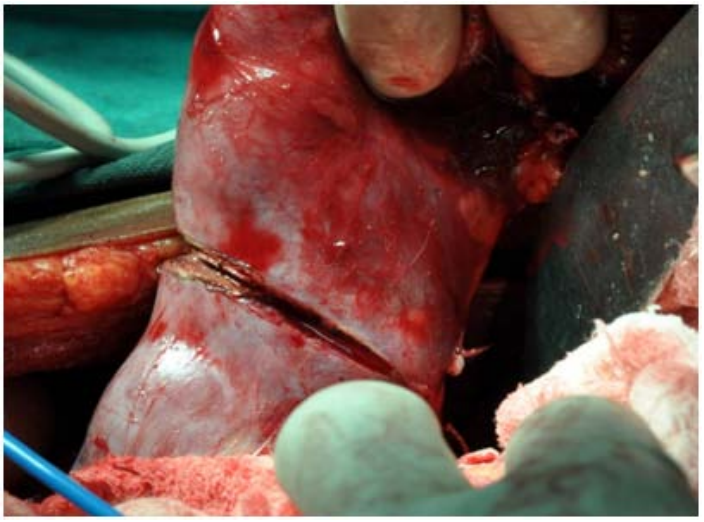
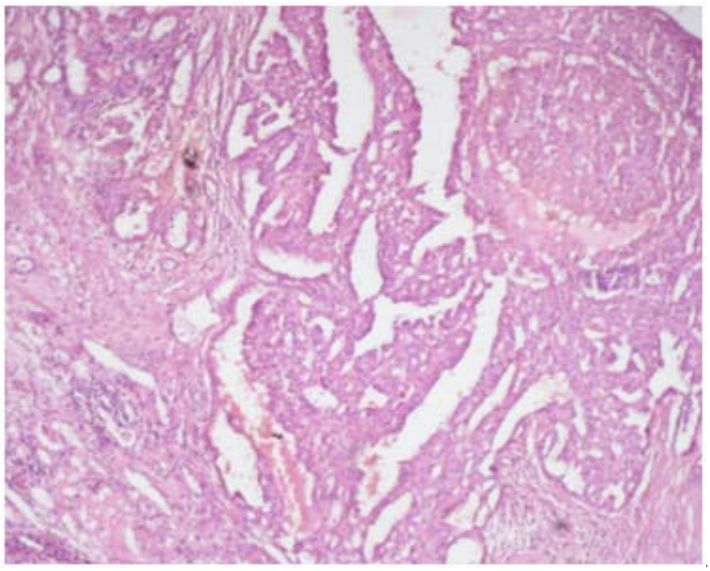


Figure 3. The microscopic appearance of type II papillary RCC demonstrating eosinophilic cells.



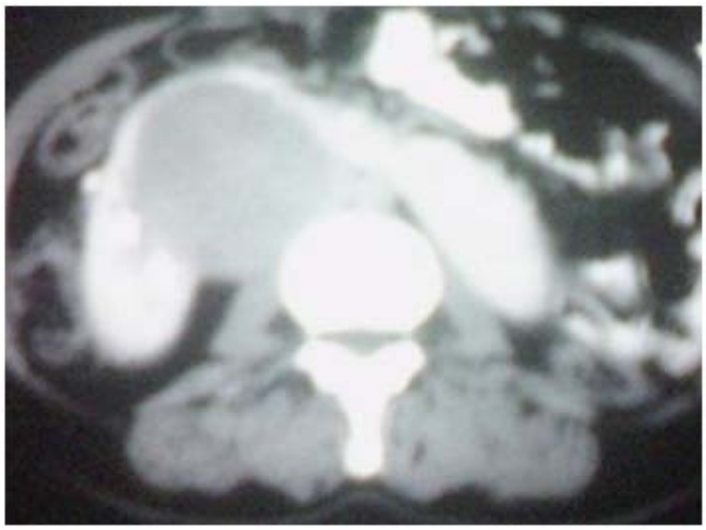
formation. Postoperative recovery was uneventful and the patient was discharged on the fourth postoperative day. The microscopic diagnosis was papillary carcinoma type II with no invasion of the capsule and negative surgical margins (Figure 3). The patient is under regular postoperative surveillance without any local recurrence or metastatic activity after 10 months postoperation.

Case Number 2

A 40-year-old female patient presented to our OPD with a chief complaint of right flank pain with a few episodes of painless, total gross hematuria with the passage of clots. There was no history of dysuria, fever, graveluria, weight loss, or anorexia. On physical examination, an 8 cm x 7 cm lump was palpable in the right lumbar region. Her baseline blood investigations were within the normal limit. An ultrasonography of the abdomen revealed HSK with a mass approximately 10 cm x 8 cm arising from the lower pole of the right renal moiety. A multidetecting CT confirmed HSK fused at the lower pole with an enhanced 10 cm x 8 cm lesion arising from the lower pole of the right renal moiety and adjacent isthmus, along with areas of calcification and necrosis suggestive of RCC (Figure 4). Metastatic work-up, including a chest X-ray and liver function test, was normal. On exploration, HSK with a mass arising from the lower pole and part of the isthmus was seen. The patient underwent transperitoneal right radical nephrectomy with isthmectomy. Intraoperative findings of a HSK with a mass arising from the right lower pole involving part of the isthmus were confirmed. No significant lymphadenopathy was noticed. The stump of isthmus was run under with absorbable running sutures, and then it was covered with an omental patch to prevent hemorrhage and urinary fistula formation. Postoperative recovery was uneventful. A microscopic diagnosis revealed the clear-cell variety of RCC with no invasion of the capsule and negative surgical margins.

DISCUSSION

Figure 4. A CT scan showing the mass arising from the lower pole of the right renal moiety and isthmus of a horseshoe kidney.



Horseshoe kidney is the most common fusion anomaly, with an incidence of 0.25% in the general population [1]. Jacopo Berengario da Carpi first described it in 1522 during an autopsy, but Botallo, in 1564, presented the first extensive description and illustration of a horseshoe kidney [5]. It is found to be more frequent in men, with a 2:1 male/female ratio. This anomaly consists of 2 distinct renal masses lying vertically on either side of the midline and connected at their respective lower poles by a parenchymatous or fibrous isthmus that crosses the midplane of the body. Embryologically, the abnormality occurs between the fourth and sixth week of gestation after the ureteric bud has entered the renal blastema. There are several variations in the basic shape of the horseshoe kidney. In 95% of cases, the kidneys join at the lower pole, which occurs before the kidneys have rotated on their long axes. In a small subset, an isthmus connects both upper poles [6].

Because of the increased use of ultrasound screening, most of the time it is diagnosed incidentally, and sometimes patients present with pictures of hydronephrosis, infection, and calculus formation. However, it is rare that a patient presents with malignancy of the renal unit. About 150 patients with horseshoe kidney presenting with malignancy of the renal unit have been reported [7]. Two cases of bilateral tumors have been reported [8]. Of all the reported cases, renal cell carcinoma accounts for about half of the cases [4] while renal pelvic tumors and Wilms tumor account for the remaining 50% (with equal incidence). Rarely, sarcoma and carcinoids have been reported. The incidence of RCC in HSK is no greater than that in the general population, but renal pelvic tumor and Wilms tumor are greater. Except for renal pelvic tumors, most of the renal malignancy in HSK arises from the isthmus [4,9]. The first case report in our case series is very rare because of the origin of RCC from the superior pole of the right renal moiety of a female patient. Moreover, the histological finding of papillary type II RCC is also unusual as most of the individual case reports in literature for RCC in HSK is of the clear-cell variety. Papillary renal cell carcinoma is typically composed of cells with abundant, clear to faintly eosinophilic, cytoplasm arranged in nests and papillary structures, as seen in Figure 3 of our case [10]. Papillary RCC is the second most frequent RCC subtype after clear-cell carcinoma accounting for approximately 13 to 15% of all known diagnoses of RCC in a normal kidney. Type II papillary RCC is less common than type I. Preoperative information about neoplastic localization, including a local extension to the surrounding structure and vasculature, is an indispensible part of the diagnostic approach to horseshoe kidney tumors, so that complete resection of the tumor can be carried out without unnecessarily removing functional tissue. When planning the operation, the possibility of growth of the tumor through the parenchymatous isthmus from one side of the kidney into the other side should be taken into account. If the tumor is present on both sides of the isthmus, surgery for both renal units is necessary. If the tumor is small and confined to the isthmus, isthmectomy is feasible [9]. Regardless of whether the procedure is radical or organ-sparing, the division of the isthmus is essential, not only to achieve complete access to the lymph nodes, but also to normalize the course of the ureter and to prevent the potential development of Rovsing syndrome [11]. Survival from these tumors is related to the pathology and the stage of the tumor at diagnosis, and not to the renal anomaly [12]. In our case series, survival seems to be good since the tumor was localized and didn't involve perinephric fat or any adjacent structure. The objective of postoperative kidney tumor surveillance is the early identification of local recurrence and metastatic activity.

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