

Adenocarcinoma in Horseshoe Kidney

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An adenocarcinoma arising in a horseshoe kidney (HK) is rare. The case of a forty five-year-old male patient, presenting with a recurrent, painless hematuria, is reported. On investigation the patient was found to have a horseshoe kidney, with an adenocarcinoma in the left hemi-kidney, which was treated surgically, with a hemi-nephrectomy, of the involved part, being performed to excise the tumor. A brief review of the relevant literature is also presented.

Key Words: Adenocarcinoma, horseshoe kidney (HK), heminephrectomy

INTRODUCTION

A horseshoe kidney (HK) is a rare, developmental anomaly that occurs in about 0.25% of the population, and arises during the 4th week of embryonic development.¹ The classical "mechanical theory" of the origin of a HK, where fusion of the medial most subdivisions of the mesonephric buds occurs, is no longer considered valid for all cases. Instead, the abnormal migration of the posterior nephrogenic area is now considered to be a more common causative factor, especially in patients with a parenchymatous isthmus.²

A HK predisposes to urinary stasis, infection and nephrolithiasis, but the development of cancer, although previously documented, is rare. A search of the available English literature revealed only 24 other cases of an adenocarcinoma arising within a HK in the last 25 years,^{1,3,5-19} taking the total of such cases to 79 till date.¹⁻¹⁸ Such a case, of an adenocarcinoma arising within a HK, is

presented herein.

CASE REPORT

A forty-five-year old male patient presented with a history of a painless hematuria of one months duration. The general physical examination was unremarkable, but examination of the abdomen revealed a hard, bimanually palpable, non-tender mass, about 8 by 6 cm in size, in the left lumbar region.

His routine hematological and biochemical investigations were within normal limits, but examination of the urine revealed a microscopic hematuria. An intravenous urogram (IVU) was suggestive of a horseshoe kidney, with adequate function, although a persistent extrinsic impression was noted in the left pelvicalyceal system (Fig. 1). A contrast enhanced computerized tomogram (CT) scan also showed a horseshoe kidney (Fig. 2). A large soft tissue mass was also noted on the left side, which was supero-medially displacing the pelvis. There was no evidence of extra-organ spread on the CT scan.

With a provisional diagnosis of a carcinoma in the horseshoe kidney, the patient was taken for surgery, following adequate preparation. The kidney was approached through a midline transperitoneal incision, and the tumor localized to the left hemi-kidney, which was seen to extend into its upper pole. A few para-aortic lymph nodes were also noted towards the hilum. The isthmus of the horseshoe kidney was divided, and a left hemi-nephrectomy was performed, taking care to ligate the supplying vessels. The para-aortic lymph nodes were also removed. After ensuring there was no urine leak from the residual kidney,

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Fig. 1. An intravenous urogram (IVU) showing a horseshoe kidney, with adequate renal function on both sides, and an extrinsic compression on the left pelvicalyceal system.



Fig. 2. Contrast enhanced CT scan of the abdomen showing a horseshoe kidney, with a large soft tissue mass on the left side. This is seen to superomedially displace the pelvis.

the procedure was completed.

The histopathology specimen weighed 1200 grams, with the cut section showing a growth of 8×7 cm in dimension. The growth had a variegated appearance towards the renal pelvis. On

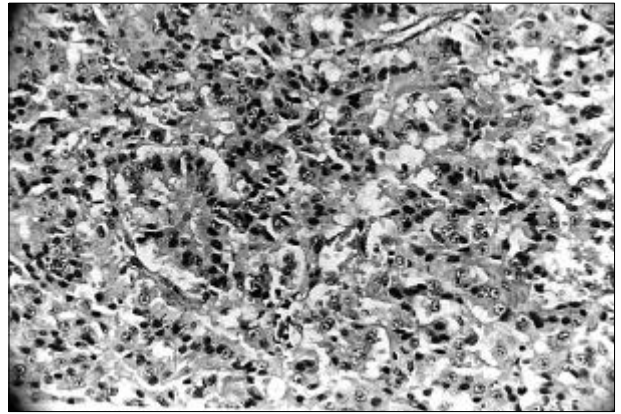


Fig. 3. Photomicrograph of the specimen showing the cellular pattern of a chromophobe eosinophilic variant type renal cell carcinoma.

microscopic examination, the cellular pattern was a chromophobe eosinophilic variant type renal cell carcinoma (Fig. 3). There was metastasis in only one of the removed para-aortic nodes. The resection margin, renal vein and ureter were free of the tumor.

The patient had an uneventful post-operative course, and was discharged on the fifth post-operative day. The patient is presently well after a one year follow up, with a normal CT scan and renal function.

DISCUSSION

A HK is often associated with other congenital anomalies of the urogenital, central nervous, gastrointestinal, cardiovascular and the locomotor systems, but tumors arising within a HK are uncommon.¹⁻³ In a major review of tumors arising within a HK, Buntley, in 1976, could only identify 55 cases of adenocarcinomas.⁴ A subsequent search of the available English literature revealed only 24 other reported cases to date.^{1-3,5-18}

An adenocarcinoma is the most common tumor to arise within a HK, but has a similar incidence in the normal population.¹⁻³ Other tumors, such as renal carcinoids, transitional cell carcinomas and nephroblastomas, have a much higher incidence in a HK than in the normal population.^{1,2} This may be a result of stasis, lithiasis and infection (in cases of transitional cell carcinoma), or due to the

abnormal migration of the posterior nephrogenic area (for nephroblastomas).^{1,2}

As always, the key to successful surgery lies in careful and good pre-operative planning. A contrast CT scan is essential in the diagnosis and management of these cases. A few authors have recommended pre-operative arteriography in these situations, which helps to identify any vascular anomalies (seen in up to 70 % of cases).^{1,2} The presence of a vascular anomaly, due to the entire HK potentially being dependent on the vessels of the isthmus, and the frequent association with hematomas or perirenal abscesses, and the possibility of the tumor extending across the isthmus and into the contra side, must always be kept in mind when surgery is indicated.^{1,2}

A midline abdominal incision provides adequate access to the entire HK and its vasculature. Surgery proceeds by carefully isolating, and ligating, the vessels to the affected part. The isthmus, as a rule, has to be divided during the removal of the cancer from the HK. Not only does division of the isthmus provide access to the draining lymph nodes, it also helps normalize the course of the remaining ureter, thereby minimizing the potential of further complications. "Reperitonealization" helps reduce the problems of bleeding and urinary fistula from the sectioned parenchyma.²

To conclude, although an adenocarcinoma is the most common tumor to arise within a HK, its incidence is no higher than in a normal kidney. Surgery entails a midline laparotomy, to facilitate exposure and gain access to the often-variable vasculature, with division of the isthmus, for adequate clearance of the regional lymph nodes, and to normalize the course of the ureters.

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