

Carcinosarcoma Of The Kidney

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Citation

J Namsupak, S Sparks, S Zaslau, S Kandzari. *Carcinosarcoma Of The Kidney*. The Internet Journal of Urology. 2007 Volume 5 Number 2.

Abstract

Carcinosarcoma of the renal pelvis is a rare malignancy of the genitourinary system. Affecting adults around the sixth to seventh decade of life this malignant tumor rapidly progresses, metastasizes, and carries a poor prognosis. Patients often present with gross hematuria and hydronephrosis of the affected kidney. Grossly and morphologically similar to sarcomatoid carcinomas, a diagnosis of true carcinosarcoma requires immunohistochemical studies. Excision of the affected organ is the only definitive treatment. Surgical management provides survival rates ranging from one month up to two years. There are few well-documented cases of true carcinosarcoma of the renal pelvis in the medical literature. We report a case of carcinosarcoma in a 74-year-old man, its clinical course and our experience in diagnosing and treating this unusual tumor.

INTRODUCTION

Carcinosarcoma of the renal pelvis is a rare, rapidly progressing malignant neoplasm with poor prognosis. In the adult population, the incidence of malignant renal sarcomas is 1% to 2% (1). Carcinosarcoma exists as a rare subtype of renal sarcoma affecting people in the sixth decade of life and beyond (1). The tumor consists of a mixture of carcinomatous and sarcomatous elements. The histologic appearance of carcinosarcoma exhibits a variety of characteristics and different histogenetic cell types (2). These characteristics contribute to the difficulty of differentiating carcinosarcoma from other tumors. Often these neoplasms are bulky, invasive, and rapidly growing. Extensive replacement of renal tissue contributes to the bulkiness of the mass and eventually hydronephrosis or hydroureter (3). The invasiveness of the tumor may be attributed to its derivation from mesenchymal stem cells (4). Commonly the presenting symptom is painless gross hematuria (5). To date fewer than 20 well-documented cases of carcinosarcoma of the renal pelvis have been reported in the medical literature.

CASE REPORT

A 74-year-old man presented with gross hematuria. Renal ultrasound showed severe left hydronephrosis. Past medical history was significant for atrial fibrillation, coronary artery disease, hypertension, and chronic renal insufficiency. Abdominal examination revealed no palpable masses. On cystoscopy the patient was found to have a 4cm papillary lesion on the anterior bladder neck and prostatic urethra. A

transurethral resection of the bladder tumor was performed. Retrograde pyelogram (RPG) was normal on the right but contrast filled only a dilated ureter on the left side. A faint poorly visualized left nephrogram was noted. Pathology of the bladder neck mass revealed high-grade transitional cell carcinoma (TCC) with squamous metaplasia without invasion to the muscularis propria or prostatic stroma. The patient underwent intravesical chemotherapy with induction BCG. Postoperative magnetic resonance imaging (MRI) of the abdomen and pelvis revealed severe left hydronephrosis and dilation of the left ureter without abdominal lymphadenopathy.

At six months, the patient had a surveillance cystoscopy and prostate biopsy for cancer monitoring. Followup MRI was performed which showed significant change in the left renal pelvis. There was now evidence of multiple filling defects on the left with an abnormal enhancing papillary like projection within the left lower pole calyx. The distal left ureter showed thickening, suggesting TCC, and multiple lymph nodes were found medial to the left hilum. Additionally, the left ureterovesicular junction had a mass like appearance. There were soft tissue mass lesions in the left ureter and adjacent retroperitoneum. Urine cytology was normal.

It was decided at this time to proceed with a left nephroureterectomy including excision of the bladder cuff and lymphadenectomy of the hilar and retroperitoneal nodes. On gross examination the patient had a severely hydronephrotic left kidney with enlarged hilar and

retroperitoneal lymph nodes. The multiloculated kidney appeared gray brown in color. Multiple grossly positive lymph nodes in the hilar region could be identified, the largest of which measured 3.5 cm. There was direct extension of the tumor into the soft tissues including the adipose tissue around the adrenal gland. However, there did not appear to be invasion of the renal vessels. The ureter lumen was completely filled with a firm friable tissue and no bladder mucosa could be appreciated. Tumor was found at the ureteral resection margin. Seven additional hilar and retroperitoneal lymph nodes were submitted of which two appeared grossly positive.

Morphologically the parts that were identified as carcinoma appeared to be an intermediate between TCC and squamous cell carcinoma. The friable tissue within the lumen of the ureter and the tissue within the hilar lymph node consist of a similar type of intermediate carcinoma. Histological examination of the kidney showed an admixture of several tissue types. The neoplasm of the renal pelvis consisted of rhabdosarcoma, osteosarcoma, and undifferentiated sarcoma components. Direct extensions into the surrounding soft tissue were also sarcomatous.

Epithelial and mesenchymal origins of the respective carcinomatous and sarcomatous elements were confirmed by immunohistochemical stains. The hilar lymph nodes and the kidney showed diffuse staining for pancytokeratin and focal staining of cytokeratin 7. However, there was no evidence of cytokeratin 20 stain. The kidney also stained strongly for vimentin. Several blocks within the kidney stained positive for desmin, S-100, and MyoD1.

One month postoperatively a cystogram was performed which showed no peritoneal leakage from the bladder. The patient subsequently underwent a course of multiagent chemotherapy and subsequently died 8 months later.

DISCUSSION

A history of unexplained onset of gross hematuria warrants radiologic investigation for a neoplasm in the urothelial organs. Few articles provide an accurate guideline regarding imaging for a carcinosarcoma of the renal pelvis. Sonography by Yilmaz et al. demonstrated a mass with a heterogenous echo pattern and areas of central attenuation. Alone sonography is not specific enough to differentiate between various malignant tumors. However, in combination with computed tomography it is possible to determine involvement of adjacent structures and vessels. Over the course of 11 months, our patient had three scans taken by

MRI. These images allowed us to track the progression of the malignancy from a slight thickening in the urothelial organs to development of new soft tissue masses and lymph node metastasis. In our experience with this case, MRI alone provided a superior imaging evaluation when compared with the combination of sonography and CT. While radiologic studies can track the progression of this neoplasm diagnosis depends on histochemical confirmation of cell origins (6).

Carcinosarcomas are theorized to arise by several mechanisms. Petersen classifies these malignant transformations into three groups. First, carcinosarcomas may develop from a pluripotent cell that differentiates along both epithelial and mesenchymal cell lines called a combination tumor. Second, concomitant malignant differentiation of a transformed epithelial cell and a mesenchymal cell within close proximity producing a single neoplasm described as a composition tumor. Finally, Petersen describes an established carcinoma that induces malignant transformation in adjacent cells. This change results in simultaneous proliferation of both epithelial and mesenchymal cell lines (7).

Two key exclusions need to be made before diagnosing a true carcinosarcoma. First, it is known that there are well-differentiated carcinomas that can transform to a less differentiated state, the transformation giving the carcinoma mesenchymal type features. Such tumors are named sarcomatoid carcinomas. Collision tumors are the second type of transformation that can be misdiagnosed as a carcinosarcoma. These tumors are formed by the fusion of an epithelial and a mesenchymal neoplasm at common borders. The two separate neoplasms appear to be one, but are still separate entities.

Using immunohistochemical stains in conjunction with morphological characteristics allowed for characterization of the neoplasm. The kidney stained positive for pancytokeratin and cytokeratin 7, which established an epithelial origin for the transformed cells. The absence of a positive cytokeratin 20 stain makes TCC unlikely. Pancytokeratin and cytokeratin staining of the hilar lymph nodes demonstrated metastasis from the renal pelvis to lymph nodes. The kidney also stained strongly for vimentin indicating tissue of mesenchymal origin. In addition, there were desmin positive cells that correlated with H&E stains of rhabdomyoblastic appearing cells. In a number of stromal cells positive for S-100 there was a small subset that stained positive for myoD1, suggesting some skeletal muscle differentiation. The positive stains for desmin, S-100, and myoD1 further

suggest carcinosarcoma by revealing tissue derived from the mesenchymal cell line.

It is important to note that only a few well-documented cases of carcinosarcoma of the renal pelvis exist. Some of these cases were reported prior to the mid 1980's and prior to immunohistochemical staining techniques. These early cases used disease progression and cell morphology to establish a diagnosis of carcinosarcoma. Using these criteria it is hard to distinguish between sarcomatoid carcinomas and true carcinosarcomas both of which rapidly progress and may appear indistinguishable on gross examination. It is highly possible that early reports of sarcomatoid carcinomas may have unknowingly been misdiagnosed as carcinosarcomas. The past two decades have given rise to widespread use of immunohistochemistry, which allows cell lines to be identified accurately despite similar appearing cell morphologies and tissue border collision. Recent reports of carcinosarcoma have included such information in order to delineate the difference between carcinosarcoma and sarcomatoid tumors. Chieh-Hsiao et al. reported a case of carcinosarcoma of the renal pelvis with a vimentin staining pattern showing an admixture similar to our findings (8).

Orsatti et al. reported a case with a disease presentation and progression analogous to the one we describe (2). Their patient was a 75-year-old man who presented with a gross hematuria and was found to have a papillary bladder tumor. After a TURBT, he had recurrent gross hematuria and was found to have a severely hydronephrotic left kidney with a tumor filled ureteral lumen. They performed a left nephroureterectomy with post-operative radiation and chemotherapy (2).

There is only one reported case of invasion of the renal vein and inferior vena cava (IVC) by a carcinosarcoma (3). Tarry reported carcinosarcoma extension into the renal vessels and IVC that was characterized by nonvisualization on intravenous pyelogram. Usually nonvisualization is attributed to either hydronephrosis or replacement of the renal parenchyma by tumor but the possibility of invasion into the renal vessels must also be considered. While we did not find invasion of the renal vessels their report illustrates an important point concerning the proper surgical approach. One must carefully image the renal vasculature and exclude tumor thrombus in the renal vein and IVC. The most successful treatment in the medical literature is

nephroureterectomy. Chen et al. reported the longest survival period which is two years (9). With the addition of postoperative chemotherapy, radiation, or combination the survival period is still unchanged (2, 5). Another treatment option to augment surgical therapy is targeted therapy with monoclonal antibodies such as those used in breast cancer (10). Further clinical study is suggested to evaluate this potential therapeutic modality.

CONCLUSION

We report a case of carcinosarcoma of the renal pelvis treated by surgical excision and multi-agent chemotherapy. This rare malignant neoplasm is aggressive and the prognosis for the patient is poor. Often the patient's only presenting symptom is hematuria. MRI is important to evaluate progression of the malignancy and metastasis. In order to distinguish a carcinosarcoma from a sarcomatoid tumor immunohistochemical studies need to be performed. The high likelihood of metastasis with this disease requires an aggressive surgical approach and consideration for chemotherapy or radiotherapy.

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