Carcinosarcoma of Kidney

Report of a Case with Emphasis on Difference from Adult Nephroblastoma

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Summary

A case of carcinosarcoma of kidney in a fifty-one year old Korean male is presented concerning with its rarity and unusual morphologic findings. Rhabdomyosarcomatous and chondrosarcomatous components comprised the bulk of tumor mass in which were proportionally abundant clear cell variety of renal cell carcinoma intermixed.

It is of authors' opinion that carcinosarcoma of kidney is very similar to adult type nephroblastoma but appears histologically different from the latter by the presence of clear cell variety predominating the carcinomatous element and by no evidence of transformation between two components.

Introduction

The concept for carcinosarcomas has been well documented since they were first described by Virchow in 1864. These neoplasms, composed of carcinoma and sarcoma inextricably intermixed, have been encountered most frequently in the uterus, breast, and esophagus, but examples of such lesions are also found in other sites, including the lungs, and nasal cavity. Only rarely have cases of carcinosarcoma primary in the kidney been reported, these have been described mostly as adult types of Wilms' tumor in literature.

Nephroblastoma or Wilms' tumor in children, also is composed of epithelial and mesenchymal components, and distinctive morphologic features of these subtypes are well recognized on the basis of differences in predominance of one or the other of these basic elements.

On the other hand, the histology and cytology of renal cell carcinoma are extremely variable, especially in the cases with a striking desmoplastic reaction (pseudosarcomatous pattern) which may also be confused with carcinosarcoma. However, they can be traced in graded stages to the characteristic polygonal neoplastic epithelial cells or may be confirmed by electron microscopy.

Other mesenchymal tumors such as sarcoma and angiomyolipomas composed of mature tissues are also seen occasionally as primary neoplasms in the kidney, but they are easily distinguished from carcinosarcomas by the absence of the epithelial component.

When these neoplasms are excluded, there remains a small group of tumors, carcinosar-
coma. But most of authors have thought that carcinomasarcoma of kidney is an adult type of nephroblastoma, although some of neoplastic components are unusual.

The purposes of this paper are as follows:
1. To report a case of carcinomasarcoma of kidney in order to summarize the morphologic peculiarities of these neoplasms by means of a review of similar cases recorded in the literature, and
2. To discuss whether it is correct to place carcinomasarcoma of kidney in the same category with nephroblastoma or not.

**Report of Case**

**History**: A fifty one-year old man was admitted to the hospital because of flank pain and abdominal mass in the left lower quadrant on September 22, 1970. He was relatively in good health until one month prior to admission, when he noticed a growing mass in his left lower quadrant of abdomen and flank pain, the latter having been more aggravated as with rapid increase of the mass for last one week before admission. Physical examination on admission revealed a moderately developed and slightly undernourished Korean male in chronic distress. A child-head sized, non-movable, relatively well outlined firm mass was palpable in the left lower abdomen. Otherwise, no abnormalities were evident. Laboratory examination showed R. B. C. of 4.2 mil/mm³, Hgb. 12gm/dl, W. B. C. 13,000/mm³. Urinalysis showed 3 positive albumin and no red cells. Simple KUB and IVP films disclosed a large ovoid mass shadow in the left abdomen, displacing intestinal loops to the right side. Chest X-ray was unremarkable. He underwent left side nephrectomy under the diagnosis of renal cell carcinoma on the next day. A child head sized tumor mass was found in the upper pole of the left kidney, accompanying no neoplastic extension either to the surrounding peritoneal adipose tissue or to the retroperitoneal organs. Left adrenal gland was displaced to the right side but free of tumor involvement.

**Pathologic Findings**: Submitted was a huge mass of tissue including left kidney, weighing 1,250 gm. in toto. The renal parenchyma was largely replaced by a large, partly lobulated, rubbery firm, ovoid tumor mass involving entire upper pole, which measured up to 12×11×10cm. The tumor appeared rather well delineated from the surrounding renal mass by the presence of grayish tan membranous fibrotic tissue but it, in part, infiltrated beyond the renal parenchymal and perinephric adipose tissues. On section it exhibited pale grayish yellow, partly lobulated solid tumor tissue with scattered intermixtures of grayish white, fleshy component and large hemorrhagic necrosis. Encountered also were multifocal, lobulated cartilaginous nodules which measured up to 5 cm. in cross. The main renal vein was full of tumor emboli with direct connection to the tumor mass, but ureteral stump remained unremarkable.

Microscopically, the neoplasm disclosed various components: the bulk comprised of loosely textured spindle shaped cells in bundles, in which were intermixed numerous bizarre acidophilic cells with occasional cross-striations indicating rhabdomyosarcomatous element and myxoid and liposarcomatous patterns in the other. Embedded within those were large neoplastic cartilaginous nests containing double nucleated lacunae and scattered mitoses, but differed from metaplastic change. The carcinomatous structures were almost identical to those of renal cell carcinoma; they consisted mainly of large clear cells in trabecular arrangement, separated clearly by thin vascular fibrous
septa. Occasionally tubular structures with dark granular cell linings were identified. Between these sarcomatous and carcinomatous elements were no areas of transformation from one to the another. None of multiple sections contained either glomeruloid or tubular structures often seen in Wilms' tumor.

Discussion

Carcinosarcoma is a rare occurrence either in childhood or among elderly people. Kim and his associates17 have found only 3 instances of these in various sites for last 15 years, each from breast, uterus and kidney from the files of Department of Pathology, College of Medicine, Seoul National University.

Three cases of carcinosarcoma of kidney were reported by Esersky and his co-workers3 in 1947 as instances of Wilms' tumor in the adult. They also reviewed the literature and included 53 prior cases. In addition to these cases, 23 cases have been found in the literature since, reported either as carcinosarcoma or adult Wilms' tumors until 1964.1 4-7, 9-17

The descriptions of these tumors and the illustrations of them leave almost no doubt that the so-called adult type Wilms' tumor and carcinosarcoma of the Kidney are closely related. The carcinomatous area, though proportionally scanty in amount, often resemble or identical with the usual forms of renal cell carcinoma, especially the clear cell and the smaller, granular cell types. Squamous cell component is a rare manifestation. The sarcomatous elements have been described chiefly as fibrosarcoma, but areas of rhabdomyosarcoma, osteogenic sarcoma, chondrosarcoma, angiosarcoma, and liposarcoma are not unusual. Multiple types of sarcomatous compositions like in the authors' case are rarely seen.

Because of histologic similarity between cases of carcinosarcoma and Wilms' tumor of the kidney, questions have been raised whether these are one the same neoplasm or not. Newman and Vellios18 stated that there is a parallel between Wilms' tumor and carcinosarcoma of the kidney, and sarcoma botryoides and carcinosarcoma of the uterus. In children, these composite tumors tend to be composed of embryonic tissue, whereas in adults the neoplasms are made chiefly of more mature types of tissues. Because of the differences which exist, they suggested that the terms sarcoma botryoides and Wilms' tumor or embryonal carcinosarcoma be reserved for the childhood types of these tumors and adult carcinosarcoma be used for the variety usually seen in adults. However, it is quite unusual to have clear cell type of epithelial component in nephroblastomas of children and adults. Chi20 reported a case of adult Wilms' tumor in which only scanty amount of tubular and glomeruloid structures of small cell linings were intermixed within rhabdomyosarcomatous and fibrosarcomatous elements. No clear cell nests were described. These evidences strongly support that carcinosarcoma of kidney is a different disease entity and can be distinguished from nephroblastoma occurring in adults, at least in the morphological point of view. When foci of transition between the carcinomatous and suspected sarcomatous elements are present, the diagnosis should be questioned19,18.

Theories of origin of carcinosarcoma may be summarized as (1) collision tumors, composed of 2 independently developing tumors that invade one another, (2) combination tumors, in which 2 different neoplastic elements are derived from 1 stem cell, and (3) composition tumors, in which epithelial and stromal cells concomitantly have become malignant. Nothing in our material can be used to support any 1
of these 3 theories. However, until more is known concerning the histogenesis of this neoplasm the term will have to be retained.

REFERENCES

14. Stout, A.P.: Human Cancer. Etiologic Factors; Precancerous Lesions; Growth; Spread; Symptoms; Diagnosis; Prognosis; Principles of Treatment. Lea & Leibiger, 1932.

Legends for Figures

Fig. 1 Cut surface of the submitted specimen. The upper pole of kidney is replaced by a huge, well circumscribed tumor tissue exhibiting multifocal hemorrhagic necrosis.

Fig. 2 Microphotograph of tumor tissue. Carcinomatous element consists entirely of clear cell variety of renal cell carcinoma in trabecular or lobular arrangement. H-E, X 100.

Fig. 3 Rhabdomyosarcomatous area of the neoplasm, comprising the bulk of tumor tissue. Tumor cells are large and bizarre containing abundant acidophilic cytoplasm. H-E, X 100.

Fig. 4 Cartilaginous nests in tumor tissue, showing many atypical chondrocytes which are often binucleated. It is adjacent to rhabdomyo sarcomatous portion with distinct border line. H-E, X 100.
Fig. 1.

Fig. 2.

Fig. 3.

Fig. 4.