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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[8]. 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[10].

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 predominence 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[8, 18]. 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-

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coma. But most of authors have thought that carcinosarcoma of kidney is an adult type of nephroblastoma[9] although some of neoplastic components are unusual.

The purposes of this paper are as follows:

1. To report a case of carcinosarcoma of kidney in order to summarize the morphologic peculiarities of these neoplasms by means of a review of similar cases recored in the literature, and

2. To discuss whether it is correct to place carcinosarcoma 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/mm^3, Hgb. 12gm/dl, W. B. C. 13,000/mm^3. 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 membraneous 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 textiled 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 allmost 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 associates[7] 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-workers[3] 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 conponent 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 Vellios[10] stated that there is a parallel between Wilms' tumor and carcinosarcoma of the kidney, and sarcoma botryoides and carcinosarcoma of the uterus. In childeren, 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. Chi[2] 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 distiguished from nephroblastoma occuring 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 questioned[10, 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 nooplastic 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.

<國文抄錄>

腎의 癌肉腫

—1症例報告 및 成人 윌름스氏 腫瘍과의 相違性에 關한 考察—

서울大學校 醫科大學 病理學敎室

金勇一·李珍鎬·文基燦·安亘煥·洪淳元·李尙國

五十一歲 韓國人 男性에 發生한 一例의 腎癌肉腫을 報告하고 이의 稀有性과 形態學的 特異性에 關하여 論議하였다.

橫紋筋肉腫 및 軟骨肉腫部位가 本 症例의 相當部分을 占有하였으나 풍부한 腎細胞癌組織도 比例的으로 混合되어 있었다.

腎의 癌肉腫은 成人型 윌름스氏 腫瘍에 매우 類似하나 腫瘍發生學的으로 獨立된 腫瘍으로 思料되었던 바, 前者는 풍부한 腎癌腫의 透明細胞型의 存在와 癌腫및 肉腫間 移行性의 根據의 缺乏等에 依하여 後者와는 組織學的으로 相異한 것이라 思料되었다.

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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. Microphotography of tumor tissue. Carcinomatous element consists entirely of clear cell variety of renal cell carcinoma in trabecular or lobular arragement. 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, X100.

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

Fig. 1.

Fig. 2.

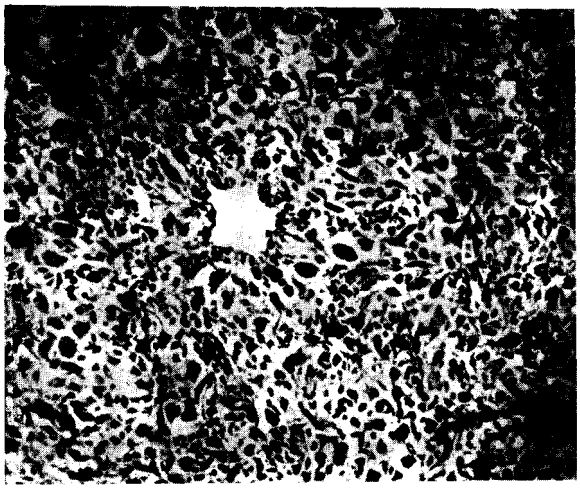

Fig. 3.

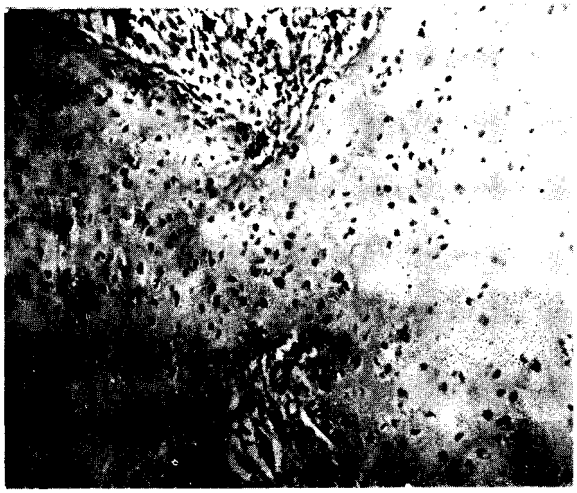

Fig. 4.