Case Report

Renal Medulla Carcinoma in a White Adolescent

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Renal medulla carcinoma is a recently described rare, aggressive tumor of the kidney occurring in patients younger than those in whom renal cell carcinoma occurs. To our knowledge, renal medulla carcinoma has been reported only in individuals of African descent carrying the sickle cell trait or afflicted by hemoglobin SC disease [1, 2]. We report what is, to our knowledge, the first case of renal medulla carcinoma occurring in a white child without a history of sickle cell disease or trait.

Case Report

A 13-year-old white girl presented with a 4-month history of right flank pain associated with weight loss and low-grade fever. Clinical examination revealed a fullness in the right upper abdomen. Sonography identified a 4.5-cm diameter cystic lesion between the right lobe of the liver and the right kidney and a second cyst within the right lobe of the liver. The right kidney was enlarged to 14.5 cm in length, with moderate hydronephrosis. An ill-defined soft-tissue mass located centrally within the kidney distorted the upper and middle calices (Fig. 1A).

A CT scan with IV contrast material showed a heterogeneous, infiltrating mass within the upper pole of the right kidney. Rim-enhancing retroperitoneal adenopathy compressed the thrombosed inferior vena cava (Figs. 1B and 1C). Sonographically guided core biopsies (using 18-gauge needles) of the renal mass revealed a mucin-producing adenocarcinoma. The patient was started on a weekly regimen of 5-fluorouracil. Subsequent CT scans of the chest and abdomen obtained 4 weeks later showed enlargement of the renal mass with new multiple pulmonary metastases. The patient died of *Candida* sepsis 1 month after the start of treatment. At autopsy, the imaging findings were confirmed, and a diagnosis of renal medulla carcinoma was made on the basis of histologic features similar to those described for this entity [1] (Fig. 2).

Discussion

Renal medulla carcinoma is an unusual, highly aggressive tumor. It has only recently been defined as a distinct pathologic entity [1]. What has been most distinctive about it has been its epidemiology. Of the 33 patients described by Davis et al. [1], all were African-Americans and all but one had sickle cell trait or hemoglobin SC disease. Consequently, an argument has been made for a genetic basis for this tumor [2]. A causal association with renal papillary necrosis, seen in the sickle cell patient group, is speculated, with a likely site of origin being within the caliceal epithelium of the renal papillae [1]. The tumor afflicts a young patient population; to our knowledge, all reported patients have been between 11 and 39 years old. The rapidity of progression of the disease is also noteworthy, with a median survival after diagnosis of only 3 months [2]. This rapid progression appears to be related to the propensity of the tumor for metastasizing early to the regional lymph nodes and the lungs. All 33 patients described by Davis et al. had extrarenal disease at the time of diagnosis, and the tumors responded poorly to current chemotherapy regimens.

The imaging findings parallel the described pathologic behavior, with the tumors located centrally within the kidney and showing an infiltrative growth pattern and associated pelvic encasement [3, 4]. In our patient, the renal mass shown on CT scanning had the typical infiltrative pattern, with heterogeneous enhancement and diffuse renal enlargement. Perinephric spread was observed, as was extension to the retroperitoneal lymph nodes. The nodes also showed the peripheral enhancement and central necrosis appearance of an aggressive process. Although the intrarenal collecting system was not visibly invaded, it was nevertheless deformed and displaced by the mass. The liver metastases were predominantly cystic. Spread to the lungs was also present.

What renders this case most unique is the occurrence of renal medulla carcinoma in a white adolescent without sickle cell disease or trait. This occurrence may necessitate revision of the genetic predisposition thesis for this condition or, at the very least, prompt the recognition of another group of patients who may also be susceptible to this aggressive neoplasm.

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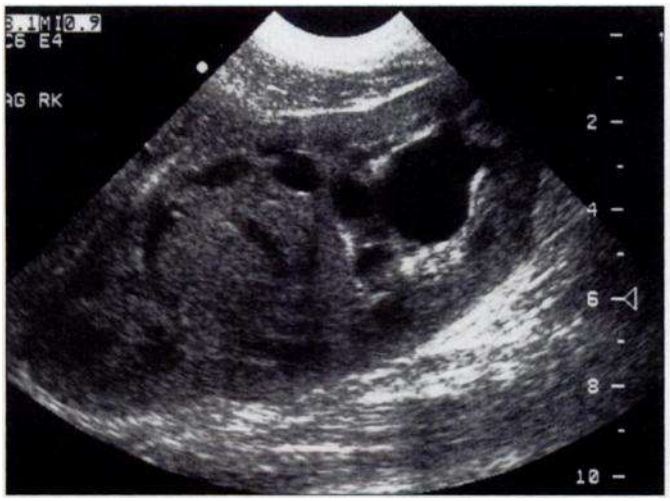

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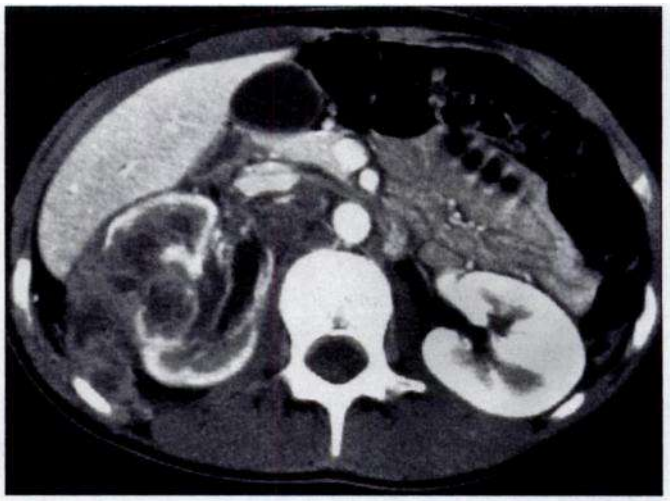

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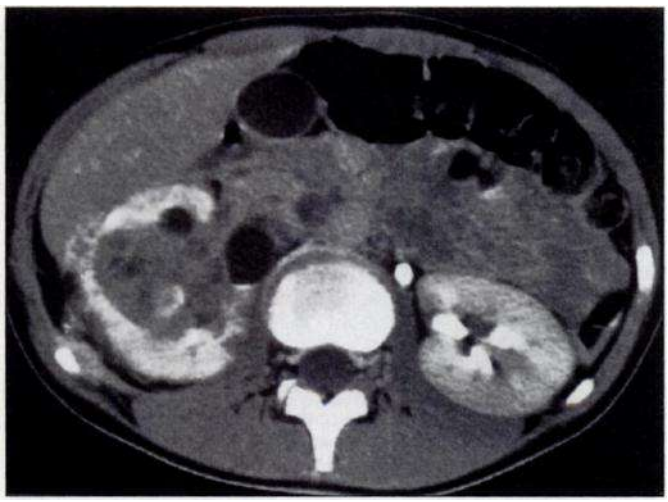

C

Fig. 1.—13-year-old white girl with renal medullary carcinoma.
A, Coronal sonogram of right kidney. Centrally located large, isoechoic mass is splaying dilated calices.
B, Helical CT scan obtained in angiographic phase shows ill-defined central mass in right kidney that extends into perirenal space. Note rim-enhancing retrocaval adenopathy.
C, Delayed CT scan obtained 10 min after **B** shows more clearly defined central renal mass.

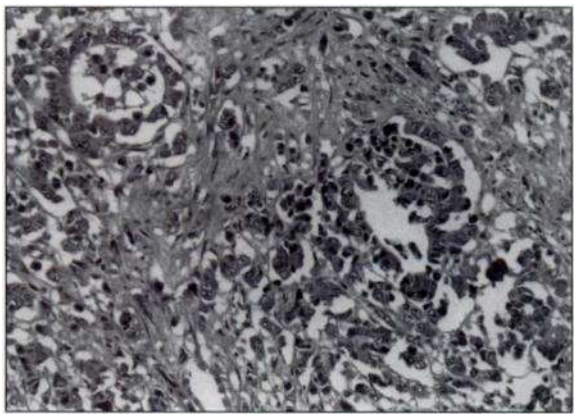

Fig. 2.—Photomicrograph showing neoplastic elements forming tubules surrounded by desmoplastic reaction. (H and E, ×200)

Acknowledgments

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