ADULT RENAL MYOEPITHELIAL HAMARTOMA

C. PETER FISCHER, M.D.
KARL R. HERWIG, M.D.
PAUL W. GIKAS, M.D.

From the Departments of Surgery (Section of Urology) and Pathology, University of Michigan Medical Center, Ann Arbor, Michigan

ABSTRACT — A renal myoepithelial hamartoma presented as a lucent filling defect with gross hematuria in an adult female. Preoperative studies caused conflicting impressions. The predominance of smooth muscle and incorporated tubuloepithelial elements characterize the tumor as a hamartoma of myoepithelial type. Pertinent review of the literature confirms the rarity of this lesion in adults.

Myoepithelial hamartoma is best described as part of a spectrum of benign renal lesions including hamartoma, leiomyoma, fibroma, lipoma, angioma, and adenoma. All occur rarely, but are particularly unusual when presenting as isolated, symptomatic lesions antemortem. Benign renal tumors when clinically significant can occur at any age. The youngest, a newborn, reported by Zuckerman et al.; the oldest, age eighty-one, reported by Bailey and Harrison. Foster, in a review of 135 cases, found an average age at presentation of thirty-nine years, a 2 to 1 female predominance, and rarity in the black race with only 2 reported cases.

Case Report

A forty-four-year-old white woman was admitted to this Center for an evaluation of gross hematuria. Two weeks prior to admission the patient experienced two episodes of total, gross, painless hematuria. Evaluation by her private physician included normal findings on physical examination and persistent microscopic hematuria without bacteriuria. Serum creatinine was 0.7 mg./100 ml., hemoglobin 13 Gm./100 ml., and the white blood cell count 6,000 cells/mm.

Bilateral selective renal arteriography and venography were performed. Aortography showed displacement of the segmental arteries and a 4-cm. radiolucent mass (Fig. 1E) in the lower part of the left kidney. The mass was avascular, and all arterial branches constricted with epinephrine infusion. Angiographic impression was that of a radiolucent avascular parapelvic cyst. Percutaneous "cyst" puncture and aspiration under local anesthesia were attempted but no fluid could be aspirated. Injected contrast media pooled around the needle tip suggestive of a solid lesion (Fig. 1F).

Because of these conflicting impressions a diagnosis of atypical hypernephroma or solid tumor of the kidney was considered. The patient underwent exploration via an anterior transperitoneal approach, and a firm area in the left lower pole was exposed via a nephrectomy. This revealed a pearly white, solid tumor.
FIGURE 1. Excretory urogram, fifteen-minute radiograph, left lower pole lucent mass is evident with distortion of lower renal pelvis and calyces (arrows). (B) Tomogram, 8 cm. cut. Mass (m) more clearly defined with left renal pelvis and lower pole calyceal distortion. (C) Prone, longitudinal, left renal sonogram shows solid mass (m) in region of lower pole. (D) Aortogram, arterial phase. There is splaying of segmental arteries around left lower pole mass (arrows), but no neovascularity. (E) Nephrographic phase of aortogram demonstrates left lower pole mass (m) to be avascular. (F) Left renal “cyst” puncture during excretory urography. Solid nature of lesion is demonstrated by pooling of injected contrast medium around needle tip.

seemingly fixed to a lower pole calyceal infundibulum (Fig. 2). Frozen section biopsy was interpreted as benign supporting tissue tumor, probable leiomyoma. The exact nature of the entire tumor was not clear, and a simple nephrectomy was performed. Postoperative recovery was unremarkable, and the patient was discharged on the sixth postoperative day.

The gross specimen consisted of a 198-Gm. kidney with a smooth cortical surface. A 3.5 by 2.7 by 2.4-cm. firm, white mass with a whorled pattern and several small cysts was noted in the medulla. The mass distorted but did not involve the calyces. Microscopically, a circumscribed but nonencapsulated nodule was present in the renal medulla. It consisted of interlacing bundles of smooth muscle, tubular structures, and cysts of variable size. A trichrome stain confirmed the presence of smooth muscle. The various components had a benign histologic appearance, and were interpreted as a myoepithelial hamartoma. The adjacent renal parenchyma exhibited no significant abnormality (Fig. 3).
Benign renal tumors have been classified and characterized by Fuchsman and Angrist who separated them into epithelial and connective tissue types. A third group described as mixed with combinations of smooth muscle, tubulopithelial elements, fibrous stroma, and adipose tissue, include the hamartoma. Hamartomas were described by Albrecht and Robbins as tumor due to overdevelopment of some tissue elements which normally belong at the site where found, or to an abnormal relationship of a normally situated tissue element. Angiomyolipoma, coined by Morgan, Straumfjord, and Hall, represents a specific type of hamartoma with the histologic features of smooth muscle, vascular, and adipose tissue in varying proportions. It falls within the broad definition of hamartoma and the two are often used interchangeably in the literature. Recent reviews by Price and Mostofi and Bissada et al. confirm the general acceptance of an embryonal origin of these tumors.

Reports of clinically significant renal hamartoma have been few. Initial reports of such a lesion were published in 1942 by Moolten in association with tuberous sclerosis — a complex characterized by epilepsy, mental retardation, adenoma sebaceum, and "tumor-like malformation in various organs," including the kidney.

Fikner, Mallory, Rusche, Fuller, and Hulse and Patik were the first to report isolated cases of symptomatic renal hamartoma without associated tuberous sclerosis. Over one hundred forty-five such cases have since been published. Presentations included flank pain, flank mass, acute retroperitoneal hemorrhage, hypertension, acute pyelonephritis, and hematuria in order of reported frequency.

Clinically, cases of symptomatic isolated hamartoma occur in the fifth to seventh decade of life with a 4 to 1 female predominance and equal distribution with respect to right and left renal involvement. They are frequently large in size, unifocal, and unilateral. When associated with tuberous sclerosis, they have an equal sex distribution; if symptomatic, they tend to present at an earlier age, are frequently small, multifocal, and bilateral. Microscopically, both isolated cases and those associated with tuberous sclerosis have been typical angiomyolipomas. The presence of tubulopithelial elements in our case is more consistent with a true hamartoma and has not previously been reported except in children, where a diagnosis of Wilms tumor was initially considered. Long-term follow-up, up to eleven years in the Price and Mostofi series, confirms the benign nature of these tumors.

When presented with a lucent filling defect on excretory urography that is solid by ultrasonography and cyst puncture, but avascular by selective angiography, the differential diagnosis must include necrotic or avascular hypernephroma, sarcoma, benign solid tumor, and polycystic kidney disease. Angiographic characteristics peculiar to hamartoma have been described. A recent review of 26 cases found only 32 per cent with one or more of the typical findings, and 68 per cent with neovascularity indistinguishable from hypernephroma. Fewer
than 5 cases have been correctly diagnosed preoperatively. Management can thus be difficult and may require exploration to resolve the issue. Although approaches to diagnosis and management are well reviewed by McCullough,17,21 the variable clinical and pathologic spectrum of isolated renal hamartoma make individualization necessary — particularly when, as in this case, sophisticated diagnostic techniques are inconclusive.

Ann Arbor, Michigan 48109
(DR. FISCHER)

References