

# Mixed Epithelial and Stromal Tumor of the Kidney

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A 42-year-old female presented with intermittent left flank pain for 2 years. She developed gross hematuria 1 month prior to seeking help. Intravenous pyelography showed filling defects within the lower portion of the left collecting system and renal pelvis causing hydronephrosis. Abdominal computed tomography revealed a huge cystic heterogenic tumor about 20 cm in largest diameter occupying the entire left kidney. A left radical nephrectomy was performed without complications. The pathology report confirmed the diagnosis of mixed epithelial and stromal tumor of the kidney. From pathologic survey, the spindle cells of this tumor were positive for muscle markers and expressed estrogen and/or progesterone receptors. We suggest that a mixed epithelial and stromal tumor of the kidney should be considered in all cystic renal tumors presenting in perimenopausal women. [*J Chin Med Assoc* 2006;69(3):140–142]

**Key Words:** mixed epithelial and stromal tumor, renal cyst, renal tumor

## Introduction

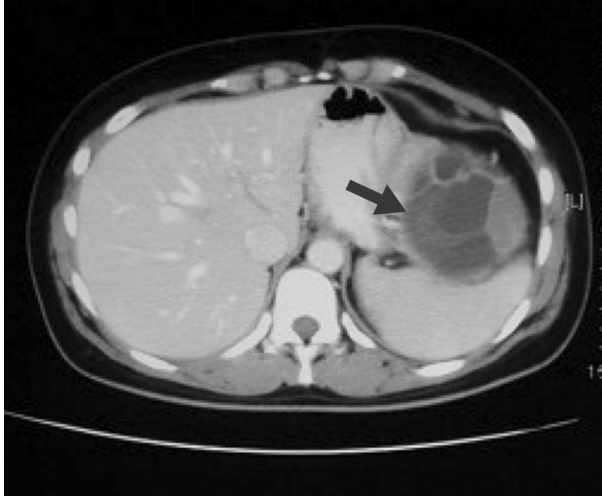
In recent years, a rare distinctive kidney tumor composed of a mixture of stroma and epithelium with solid and cystic architecture has been recognized.<sup>1–5</sup> These tumors, published earlier under different nomenclatures such as “adult type of mesoblastic nephroma”, “cystic hamartoma of the pelvis”, “adult type of cystic nephroma”, “leiomyomatous hamartoma”, and “solid and cystic biphasic tumor of the kidney”, represent, in fact, the same neoplasm, with a spectrum of differentiation.<sup>3</sup> The name “mixed epithelial and stromal tumor of the kidney” was first introduced by Michal and Syrucek in 1998.<sup>5</sup> Here, we present a case of this entity, found accidentally.

## Case Report

A 42-year female patient had suffered from intermittent left flank pain for 2 years before she developed gross hematuria in January 2004. She visited our outpatient department for help. She denied any hormonal drug or surgical history. She was not married and denied

gestation history. No abnormal physical examination result was noted, except mild left flank knocking pain. Routine urine analysis revealed microscopic hematuria (red blood cells, 3–4 per high-power field). Urine cytology was negative for malignancy. Intravenous pyelography showed filling defects within the lower portion of the left collecting system and renal pelvis, causing hydronephrosis. Abdominal computed tomography revealed a huge cystic heterogenic tumor about 20 cm in largest diameter occupying the entire left kidney (Figure 1). Left radical nephrectomy was undertaken in February 2004. The pathology report surprisingly showed a “mixed epithelial and stromal tumor of the kidney”, with progesterone receptor (+), estrogen receptor (+), AE1/AE3 (+) (for epithelial cells), desmin (focal +), and vimentin (focal +) (Figure 2). However, the preoperative hormone profile of the patient was not available. From a hormone survey 1 month postoperatively, the progesterone level was 0.73 ng/mL and the estrogen level was 65.4 pg/mL, both in the normal range. The patient has received regular follow-up at our out-patient department. No evidence of tumor recurrence has been found for 14 months.

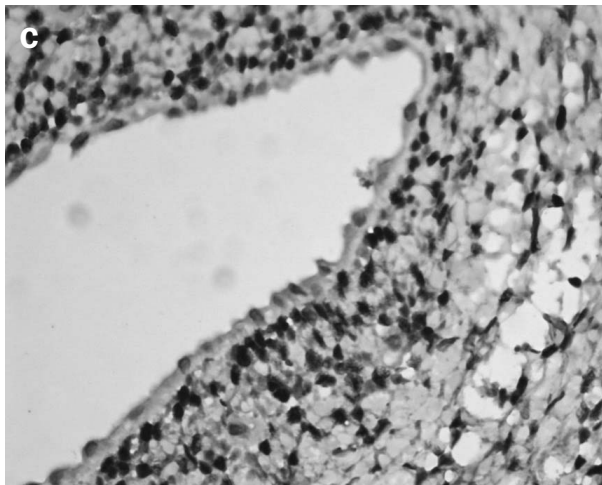
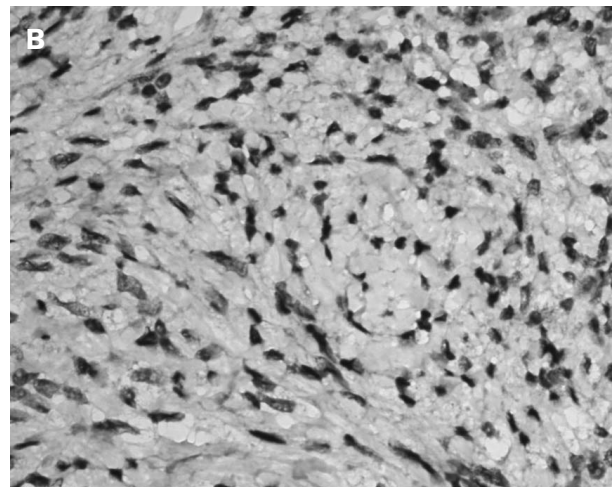
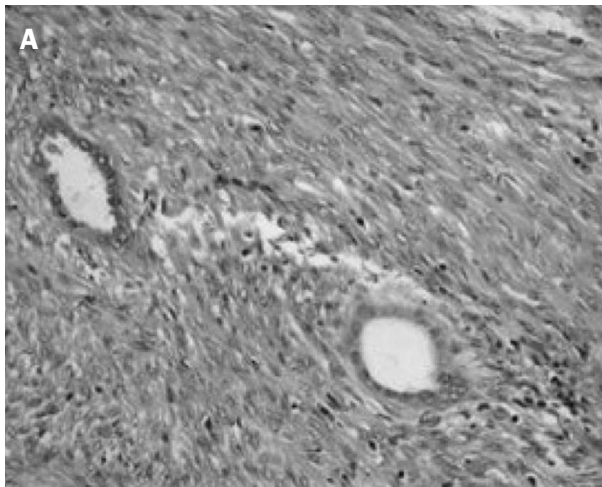
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**Figure 1.** Abdominal CT scan revealed a left renal tumor with cystic and solid components (arrow).

## Discussion

Up to June 2000, 12 cases of mixed epithelial and stromal tumor of the kidney have been reported in North America by Adsay et al.<sup>1</sup> Another 38 probable cases have been reported worldwide.<sup>1</sup> From the study of Adsay et al, most patients with this tumor present with flank pain, hematuria, or symptoms related to genitourinary tract infections, and some are asymptomatic, detected incidentally during investigation for other diseases.<sup>1,2</sup> They also found a hormonal relationship among these patients.<sup>1,3,4,6-8</sup> That is, most female patients, but not all, had a history of treatment with estrogen–progesterone or had undergone prior gynecologic surgery. The only male patient had a long history of diethylstilbestrol therapy. Additionally, all female patients were around



**Figure 2.** (A) Pathologic examination showed tubular glandular epithelium scattered within abundant spindle cells (hematoxylin and eosin,  $\times 300$ ). (B) Immunostains showed estrogen receptor (+). (C) Immunostains showed progesterone receptor (+) with brownish discoloration.

the perimenopausal age.<sup>1,2</sup> In the pathologic survey, the epithelial component of this tumor consists of glands with different complexity and distribution, and the stromal component is characterized by a spindle cell proliferation. The spindle cells of this tumor are frequently positive for muscle markers and often express estrogen and/or progesterone receptors.

In the literature, it is postulated that a deranged hormonal milieu (perimenopausal changes or therapeutic hormones with unopposed estrogen) induces the proliferation of “peri-ductal fetal mesenchyme”, which has the capacity for dual (mesenchymal and epithelial) differentiation and presents around the epithelial structures in organs like the kidney, liver, and pancreas.<sup>1,2</sup> The prognosis of this tumor is usually very favorable. Only 1 malignant case was reported recently.<sup>3,5</sup>

In comparing our case to others, we found several different points. Our patient is younger and not menopausal, in contrast to the mean age of 56 years reported by Adsay et al.<sup>1,2</sup> Our patient did not have any hormonal drug or prior gynecologic surgery history. This is quite different from other studies. There seems to be little hormonal influence in the tumorigenesis of our patient. However, the expression of spindle cells in our patient was positive for estrogen and progesterone receptors. This is the only fact that correlates well with the hormonal theory raised by others. Other possible hormonal factors in our patient warrant further investigation.

Mixed epithelial and stromal tumor of the kidney usually presents in perimenopausal women as a partially cystic mass, and its growth may be influenced by hormones. Any cystic tumor presenting in perimenopausal women should undergo the same series of tumor surveys as for malignant tumors. A mixed epithelial and stromal tumor of the kidney should be considered one of the possibilities.

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