

# Peritoneal seeding following incomplete resection of mixed epithelial stromal tumor of the kidney: First case report

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## Abstract

Mixed epithelial stromal tumor (MEST) is rare and typically benign renal cystic neoplasm that cannot be clinically distinguished from cystic renal cell carcinoma. Its mainstay course of diagnosis and treatment remains surgical excision. Recurrence and malignant transformation is rare but has previously been described. To our best knowledge, we present the first case of peritoneal seeding resulting in a paracolonc MEST following incomplete resection in a patient with benign MEST. This signifies a new pathological behavior for MEST, predominantly, a benign kidney tumor. In addition, documentation with more cases of MEST is needed to further understand its pathogenesis, clinical behavior, malignant potential, and optimal management.

**Key Words:** Adult mesoblastic nephroma, benign kidney tumor, benign renal cyst, cystic nephroma, mixed epithelial stroma tumor, mixed epithelial stromal tumor, paracolonc cyst, recurrence

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## INTRODUCTION

Mixed Epithelial Stromal Tumor (MEST) is a rare and typically benign renal cystic neoplasm. It was first described as its own entity by Michal *et al.* in 1998, differing from other renal tumors of epithelial and mesenchymal differentiation.<sup>[1]</sup> Histologically, it is a distinctive tumor characterized by its biphasic pattern composed of varied epithelial elements that form glands and cysts embedded within proliferative spindle cell, ovarian-like stroma staining positive for estrogen and progesterone receptors.<sup>[2]</sup> Previous terminology includes adult mesoblastic nephroma, leiomyomatous renal hamartoma, solid and cystic biphasic tumor, cystic hamartoma, and adult metanephric stromal tumor.<sup>[3]</sup> Classic presentation is that of a middle aged

peri-menopausal female with history of long term estrogen therapy. Macroscopic features include a mass with solid and cystic components, usually centrally located that may herniate into the pelicalyceal collecting system. On CT imaging, it is a well-circumscribed multiseptate cystic and solid mass with delayed contrast material enhancement often categorized as a Bozniak III or IV cyst.<sup>[4]</sup> The differential diagnosis is extensive and includes angiomyolipoma with cysts, synovial sarcoma of kidney, sarcomatoid carcinoma, leiomyosarcoma, multicystic dysplastic kidney, obstructed duplicated renal system, renal abscess, and multilocular cystic renal cell carcinoma (RCC).<sup>[4,5]</sup> Since MEST cannot be clinically distinguished from cystic renal cell carcinoma, surgical excision is the mainstay course for

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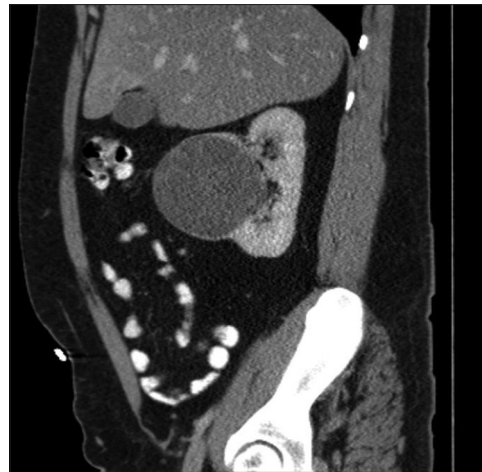
definitive diagnosis and treatment. Although considered a benign tumor, MEST can have malignant transformation with recurrence as previously reported.<sup>[6-10]</sup> To our best knowledge, we present the first case of peritoneal seeding resulting in a paracolic MEST following incomplete resection in a patient with benign MEST.

## CASE REPORT

The patient is a 38-year-old Caucasian female, who presented with right flank pain. She denied any gross hematuria or history of urinary tract infections. Her past medical history was significant for hypertension and polycystic ovarian syndrome on metformin. She was a nonsmoker and had no family history of genitourinary diseases. She had no history of hormonal therapy. A computed tomography (CT) scan of the abdomen and pelvis renal mass protocol showed a 9 cm mass with fluid density and very thin intracystic septations representing a Bosniak II cyst in her right kidney [Figure 1]. Due to flank pain resulting in multiple visits to the emergency room, the patient elected surgical treatment. She underwent robotic-assisted renal cyst marsupialization via transperitoneal approach and tolerated the procedure well with an uneventful postoperative course. Her surgical pathology showed mixed epithelial stromal tumor (MEST) measuring 6 cm in maximum dimension and focally present at the cauterized edge. She was seen at a 3-week postoperative visit. Although she had a positive margin, the plan was to return in 1 year with repeat imaging owing to the benign nature of this disease. However, she was lost to follow-up and presented 3 years later with a CT scan showing an 11 cm × 8 cm right renal cystic lesion with few thin internal septations and focal punctate calcifications along the walls of the cyst. Interestingly, she was also found to have multiple cystic lesions near the ascending colon with similar radiologic features as the renal cyst suspicious for MEST deposits [Figure 2]. She was referred to colorectal surgery, and a colonoscopy was performed that did not show any intraluminal abnormalities. Due to recurrence and fear of malignancy, the patient underwent a hand-assisted laparoscopic radical nephrectomy by urology team. Then she underwent an excision of cystic multiloculated mesenteric lesion that was attached to the ascending colon [Figure 3] by the colorectal team using Endo GIA™ stapler. Final surgical pathology showed MEST in the right kidney and also in the paracolic mass. The patient was discharged on postoperative day 2 with no issues.

## Pathology

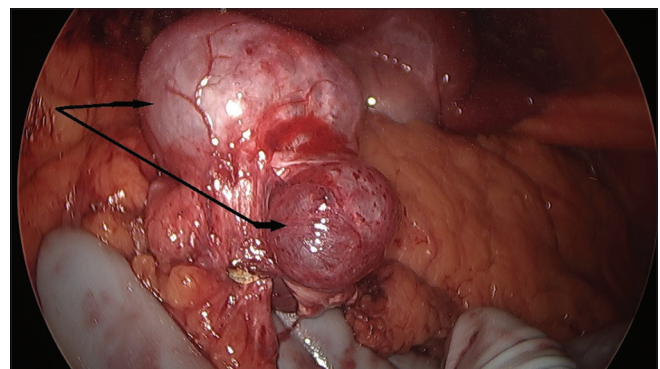
The multiloculated cystic lesion showed epithelial and stromal components. Most cysts were lined by flat to cuboidal and in some areas with hobnailed epithelium. No atypia is present in the epithelial component. The stroma was composed of



**Figure 1:** Initial 9 cm right renal cyst

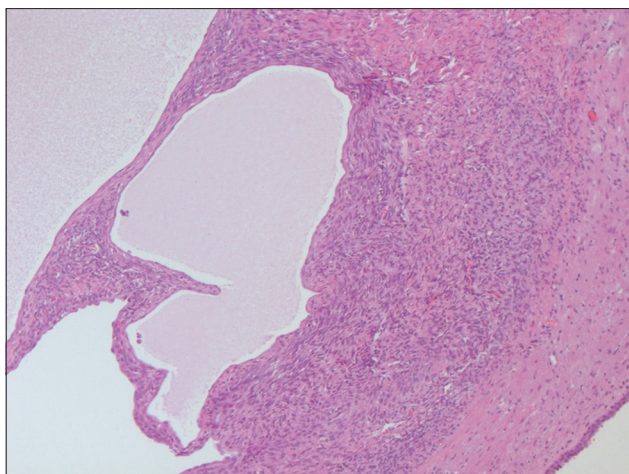


**Figure 2:** Paracolic cystic lesion (white arrow) along the ascending colon in relation to recurrent right renal mixed epithelial stromal tumor cyst (red arrow)



**Figure 3:** Mesenteric multiloculated paracolic mixed epithelial stromal tumor lesion (arrows)

areas of dense fibrous, loose, smooth muscle, hypercellular spindle, and ovarian stroma-like elements [Figure 4]. No mitotic activity or atypia was observed in those stromal components either. The septae did not contain any normal renal parenchyma.



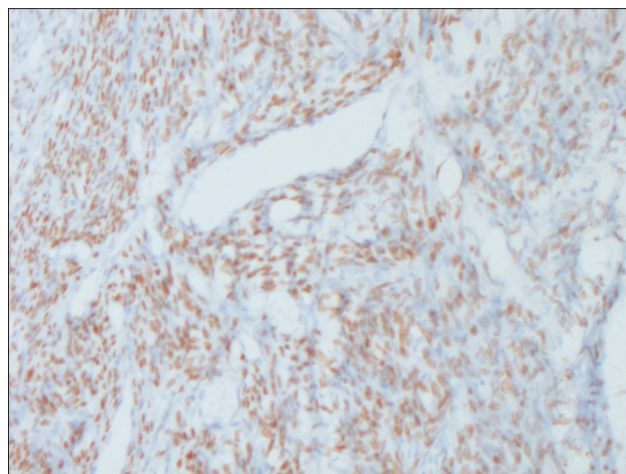
**Figure 4:** H and E stain of the renal cyst showing dense fibrous, loose, smooth muscle, hypercellular spindle, and ovarian stroma-like elements (x40)

### Immunohistochemistry

Stromal cells showed diffuse nuclear estrogen and progesterone positivity, consistent with MEST or also known as cystic nephroma [Figure 5].

### DISCUSSION

There appears to be approximately around 100 cases of MEST reported in the literature.<sup>[11]</sup> Due to its rare occurrence and recent recognition as a distinct tumor, it is not very well understood. A hormonal mechanism for MEST's pathogenesis has been postulated secondary to the female predilection, association with hormonal use, and estrogen/progesterone positivity.<sup>[2]</sup> Pathologically, the differential diagnosis includes multilocular cystic renal cell carcinoma, tubulocystic carcinoma, cystic partially differentiated nephroblastoma, and metanephric adenofibroma. Clinically, it cannot be distinguished from other renal cystic lesions and should be considered as high on the differential diagnosis in a middle-aged female patient with a history of hormone therapy. A nephron sparing approach is encouraged even for large masses compressing the collecting system.<sup>[12]</sup> In our patient, we did not obtain a complete negative margin at the time of her initial surgery. Owing to the perceived benign nature of MEST, we elected for active surveillance with periodic imaging. However, due to the rapid rate of her recurrence and fear of malignancy, we elected to perform radical nephrectomy and excision of the paracolic mass. Recently, a similar case has been reported in the literature in a female patient treated with robotic decortication for a complex cystic mass that recurred 2 years later, which was then treated with nephrectomy and found to have benign MEST.<sup>[13]</sup> To our best knowledge, we present the first case of local recurrence with benign MEST of kidney and peritoneal seeding resulting in a paracolic MEST. Seeding of cells into the peritoneal cavity following incomplete resection represents a new pathological



**Figure 5:** Immunohistochemical stain of stroma is positive for estrogen receptor (x200)

behavior for MEST, predominantly a benign kidney tumor. In addition, documentation with more cases of MEST is needed to further understand its pathogenesis, clinical behavior, malignant potential, and optimal management.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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