Acta Radiologica Open
11(5) 1–4

DOI: 10.1177/20584601221103019
journals.sagepub.com/home/arr
SAGE

Mixed epithelial and stromal tumor of the kidney composed mainly of solid components: A case report

Kiyoka Maeba[1], Akihiko Kanki[1], Hiroyuki Watanabe[1], Akira Yamamoto[1], Yasuto Fujimoto[2], Miyaji Yoshiyuki[3] and Tsutomu Tamada[1]

Abstract

Mixed epithelial and stromal tumor (MEST) is a relatively rare lesion of mixed epithelial and mesenchymal origin, consisting of epithelial components that form cysts and stromal cells that are positive for estrogen and progesterone receptors. The present case was a 54-year-old female who presented with hematuria. Abdominal ultrasonography revealed a 41 x 30 mm tumor in the right kidney, with the tumor protruding outward in the direction of the renal pelvis. Dynamic contrast-enhanced computed tomography and magnetic resonance imaging confirmed a solid tumor in the right kidney that showed gradual contrast enhancement and contained a central non-enhancing area with the appearance of a cystic component. Based on the imaging findings, the provisional diagnosis was papillary renal cell carcinoma or angiomyolipoma with epithelial cysts. Right nephrectomy was performed and the tumor was confirmed histopathologically as MEST. We report a very rare case of MEST that was composed mainly of solid components.

Keywords

Kidney, mixed epithelial and stromal tumor, computed tomography, magnetic resonance imaging, solid component

Received 27 November 2021; Accepted 10 May 2022

Introduction

Mixed epithelial and stromal tumor (MEST) is an epithelial–mesenchymal mixed tumor that was first reported by Michal and Syrucek in 1998.[1] It comprises epithelial components that form cysts and stromal cells that are positive for estrogen and progesterone receptors.[1-3] Prior to 1998, the tumor was variously termed multilocular cyst with ovarian stroma, cystic hamartoma of renal pelvis, and adult mesoblastic nephroma.[2-4] The MEST family of tumors, including adult cystic nephroma, was added to the WHO 2016 classification.[5] MEST is a rare renal tumor that accounts for only 0.2% of all renal tumors, with around 100 cases reported.[6] It occurs mainly in pre- and post-menopausal women, with very few male cases, and a male–female ratio of 1:10. Involvement of long-term estrogen replacement therapy has been suggested in its development.[7] Clinical symptoms in children generally include a palpable asymptomatic abdominal mass, whereas abdominal pain with hematuria is more frequent in adults.[3,6,8] There are various tumor morphologies in the MEST family, ranging from mainly cystic to mainly solid lesion components;[5] however, most previous studies have reported imaging findings of mainly cystic components. Here, we report a very rare case of MEST that was composed mainly of solid components.

[1]Departments of Radiology, Kawasaki Medical School, Kurashiki, Japan
[2]Department of Pathology, Kawasaki Medical School, Kurashiki, Japan
[3]Department of Urology, Kawasaki Medical School, Kurashiki, Japan

Corresponding author:
Kiyoka Maeba, Radiology, Kawasaki Medical School, 577 Matsushima, Kurashiki 701-0192, Japan.
Email: kiyo_0807_0329@yahoo.co.jp

Case

A 54-year-old female presented with hematuria. She had a medical history of hysterectomy due to uterine myoma and no history of hormone treatment. All her routine blood values were within normal ranges. Abdominal ultrasonography showed a 41 x 30 mm solid mass arising from the right kidney. Multiphasic computed tomography (CT) revealed a well-circumscribed solid mass in the right kidney, with the tumor protruding outward in the direction of the renal pelvis. The unenhanced CT exhibited the tumor as slightly high density lesion. The tumor contained mainly solid components that showed weak and gradual contrast enhancement in the corticomedullary to excretion phases, as well as a small non-enhancing area (Figure 1). MRI revealed a hypointense lesion on T2-weighted imaging (WI) with no apparent capsule. The area that did not enhance on CT showed high signal on T2WI and on heavily T2WI, consistent with a cystic component. No obvious abnormal hyperintensity was observed on diffusion-weighted imaging. There was no signal change observed with chemical shift imaging, no obvious fat component, and no hemosiderin deposition (Figure 2). There was no evidence of lymph node involvement or distant metastases. Based on the imaging findings, papillary renal cell carcinoma or angiomyolipoma (AML) with epithelial cysts (AMLEC) was suspected, and right nephrectomy was performed. Macroscopically, the right renal mass was white and solid, and well circumscribed with a maximum cut surface of 41 × 30 mm. There was a cystic structure centrally. Microscopically, the tumor was dominated by collagen fibers and spindle-shaped cells with poor atypia, and contained sparse cysts and ducts of various sizes (Figure 3). Smooth muscle-like cells and adipocytes were found at some sites. Immunostaining was positive for estrogen and progesterone receptors in the stroma. In addition, the muscle marker α-SMA was diffusely positive. There was also some caldesmon, desmin, WT-1, and CD34-positive cells. HMB 45 was negative. Based on these findings, the right renal tumor was pathologically diagnosed as MEST. At 2 years postoperatively, the patient has shown no sign of local recurrence.

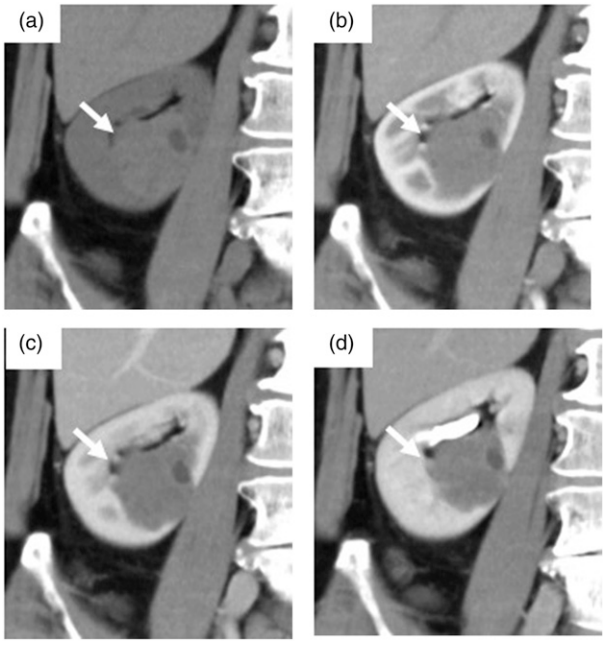

Figure 1. Multiphasic (MP) CT images in the pre-contrast (a), corticomedullary (b), nephrographic (c), and excretory (d) phases. A mass lesion is seen in the right kidney (arrow, 38 x 34 mm). The pre-contrast image exhibits the tumor as slightly high density lesion (a). It shows gradual contrast enhancement from the corticomedullary (b) to excretory phases (d). There is an additional small area that shows no enhancement on any post-contrast phase (arrowhead).

Discussion

In this case, the tumor comprised mainly solid components, and contained a small cystic component centrally. The characteristic imaging finding of MEST is of a septated cystic lesion with varying degrees of solid component.[6] Most previous reports of the imaging findings of MEST are of adult cystic nephropathy, with few reports of MEST comprising mainly solid components. However, Caliò et al. has reported the histological and immunohistological features of 53 cases of MEST, 14 of which were histopathologically predominantly solid tumors.[4] The differential diagnosis of MEST containing mainly solid components includes papillary renal cell carcinoma (RCC) and AMLEC. Papillary RCC is the second most common type of RCC, accounting for 18.5% of cases. Imaging features of papillary RCC include pseudocapsule and hypointensity on T2WI due to bleeding, iron deposition, fibrous components, and hypovascularity.[9-12] In addition, the imaging characteristic of weaker signal on in-phase than opposed-phase images due to hemosiderin deposition has been reported in papillary RCC.[10,13] The imaging findings of the present case that are consistent with papillary RCC are slightly high density on unenhanced CT and hypointensity on T2WI. However, the present lesion showed no hemosiderin deposition on chemical shift imaging and outward growth in the direction of the renal pelvis, which are inconsistent with papillary RCC. AMLEC was first recognized by Fine et al. in 2006 as a very rare subtype of AML with a cystic component, and is classified as fat-poor AML.[14] AMLEC is composed mainly of smooth muscle with very little fat, and exhibits high density on unenhanced CT, hypointensity on T2WI, and uniform contrast enhancement similar to hyperattenuating AML.[15] An imaging feature is the presence of a cyst in contact with the solid area. Imaging findings of the present case that are consistent with AMLEC are the slightly high density on unenhanced CT, hypointensity on T2WI, and central cystic component.

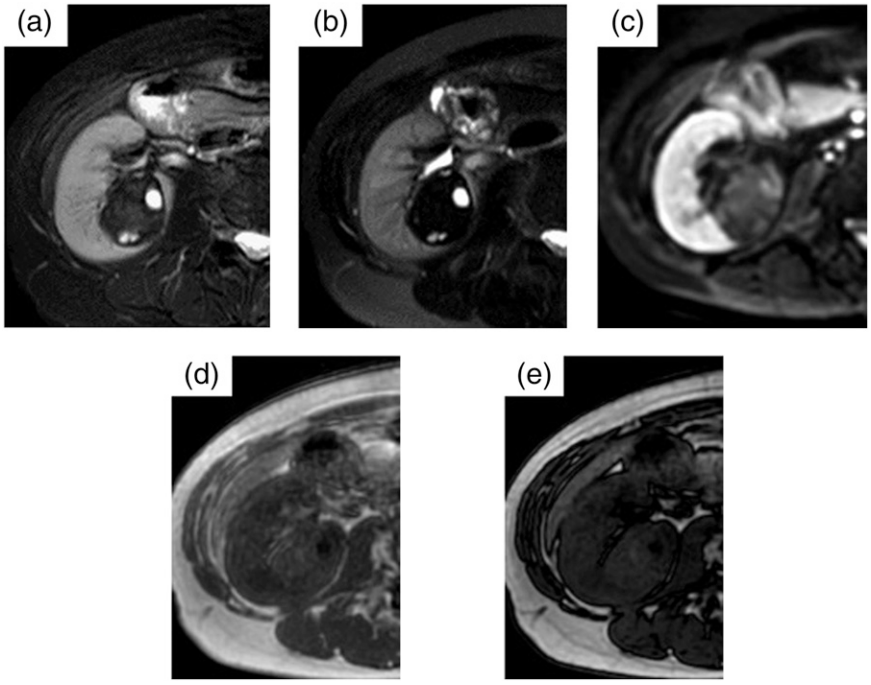

Figure 2. Magnetic resonance imaging (MRI) showed low signal mass (arrow) with tiny cyst (arrow head) on T2WI (a) and heavily T2WI (b). DWI with b value of 1000 s/mm^2 did not show hyperintensity (arrow) (c). On chemical shift imaging using in phase (d) and opposed phase (e), no signal change observed.

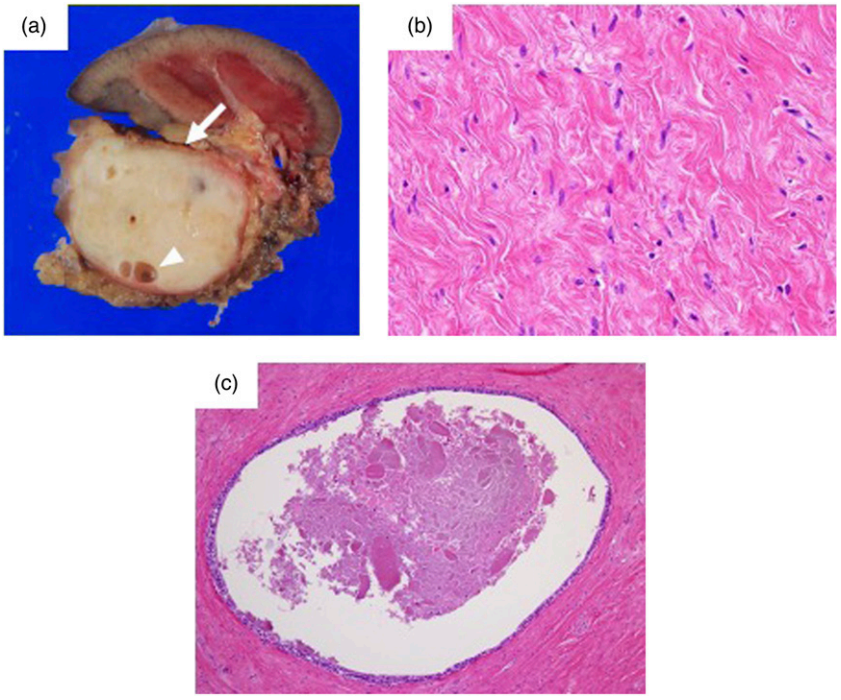

Figure 3. Gross pathologic specimen showed that the tumor (arrow) was a solid with tiny cyst (arrow head), whitish mass (a). Hematoxylin and eosin-stained images showed that the tumor mainly composed spindle-shaped cells with poor atypia and collagen fibers (b). The lining of the cystic wall was composed of flat epithelium (c). b, x400; c, x100.

Findings inconsistent with AMLEC are the outward growth in the direction of the renal pelvis and the weak and gradual enhancement pattern. Previously reported imaging features of MEST are gradual enhancement pattern of the solid component and hypointensity on T2WI due to collagen fibers.[3,16] MEST originates most commonly in the renal medulla, develops exogenously, and may invade the renal pelvis. Calcification, fat, bleeding, or capsule may be present. External development in the direction of the renal pelvis may be an important feature for distinguishing MEST from other lesions.[13,15,16]

In conclusion, MEST comprising mainly solid components is very rare. However, a diagnosis of MEST should be considered in pre- and post-menopausal patients who have a renal mass with a solid component that develops exogenously and presents as hypointensity on T2WI.

Acknowledgments

We wish to thank Takuya Moriya for his general supports.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

ORCID iDs

Kiyoka Maeba https://orcid.org/0000-0003-4196-0844
Akihiko Kanki https://orcid.org/0000-0002-2458-5598
Tsutomu Tamada https://orcid.org/0000-0003-2521-9570

References

1. Michal M, Syrucek M. Benign mixed epithelial and stromal tumor of the kidney. Pathol Res Pract 1998;194:445–448. DOI: 10.1016/s0344-0338(98)80038-1
2. Sahni VA, Mortele KJ, Glickman J, et al. Mixed epithelial and stromal tumour of the kidney: imaging features. BJU Int 2010; 105:932–939. DOI: 10.1111/j.1464-410X.2009.08918.x
3. Chu LC, Hruban RH, Horton KM, et al. Mixed epithelial and stromal tumor of the kidney: radiologic-pathologic correlation. Radiographics 2010;30:1541–1551. DOI: 10.1148/rg.306105503
4. Calio A, Eble JN, Grignon DJ, et al. Mixed epithelial and stromal tumor of the kidney: a clinicopathologic study of 53 cases. Am J Surg Pathol 2016;40:1538–1549. DOI: 10.1097/pas.0000000000000733
5. Moch H, Cubilla AL, Humphrey PA, et al. The 2016 who classification of tumours of the urinary system and male genital organs-part a: renal, penile, and testicular tumours. Eur Urol 2016;70:93–105. DOI: 10.1016/j.eururo.2016.02.029
6. Wood CG 3rd, Stromberg LJ 3rd, Harmath CB, et al. CT and MR imaging for evaluation of cystic renal lesions and diseases. Radiographics 2015;35:125–141. DOI: 10.1148/rg.351130016
7. Mohanty SK, Parwani AV. Mixed epithelial and stromal tumors of the kidney: an overview. Arch Pathol Lab Med 2009; 133:1483–1486. DOI: 10.1043/1543-2165-133.9.1483
8. Raman SP, Hruban RH, Fishman EK. Beyond renal cell carcinoma: rare and unusual renal masses. Abdom Imaging 2012;37:873–884. DOI: 10.1007/s00261-012-9903-5
9. Lee-Felker SA, Felker ER, Tan N, et al. Qualitative and quantitative MDCT features for differentiating clear cell renal cell carcinoma from other solid renal cortical masses. AJR Am J Roentgenol 2014;203:W516–W524. DOI: 10.2214/ajr.14.12460
10. Yoshimitsu K, Kakihara D, Irie H, et al. Papillary renal carcinoma: diagnostic approach by chemical shift gradient-echo and echo-planar MR imaging. J Magn Reson Imaging 2006;23:339–344. DOI: 10.1002/jmri.20509
11. Granter SR, Perez-Atayde AR, Renshaw AA. Cytologic analysis of papillary renal cell carcinoma. Cancer 1998;84: 303–308. DOI: 10.1002/(sici)1097-0142(19981025)84:5<303::aid-cncr6>3.0.co;2-7
12. Allen BC, Tirman P, Jennings Clingan M, et al. Characterizing solid renal neoplasms with MRI in adults. Abdom Imaging 2014;39:358–387. DOI: 10.1007/s00261-014-0074-4
13. Doshi AM, Ream JM, Kierans AS, et al. Use of MRI in differentiation of papillary renal cell carcinoma subtypes: qualitative and quantitative analysis. AJR Am J Roentgenol 2016;206:566–572. DOI: 10.2214/ajr.15.15004
14. Fine SW, Reuter VE, Epstein JI, et al. Angiomyolipoma with epithelial cysts (AMLEC): a distinct cystic variant of angiomyolipoma. Am J Surg Pathol 2006;30:593–599. DOI: 10.1097/01.pas.0000194298.19839.b4
15. Park BK. Renal angiomyolipoma based on new classification: how to differentiate it from renal cell carcinoma. AJR Am J Roentgenol 2019;212:582–588. DOI: 10.2214/ajr.18.20408
16. Freire M, Remer EM. Clinical and radiologic features of cystic renal masses. AJR Am J Roentgenol 2009;192: 1367–1372. DOI: 10.2214/ajr.08.1468