

**Mixed Epithelial and Stromal Tumor Family of Kidney: Clinical and MRI Features
including Application of Bosniak Classification**

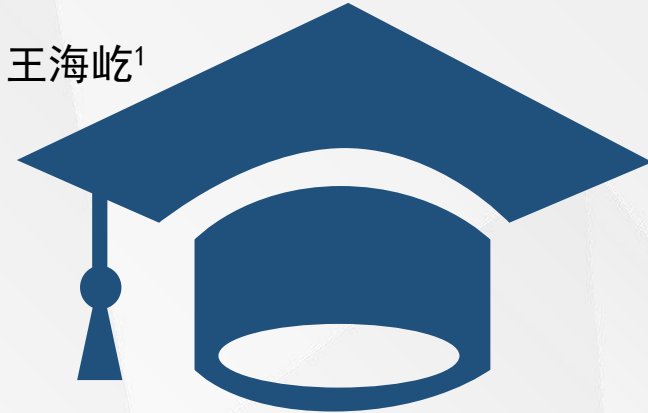
肾脏混合性上皮间质肿瘤家族的临床特点及基于Bosniak分类的MRI特点分析

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01 研究背景

- ◆ 肾脏混合性上皮间质肿瘤家族（MEST家族）是一组罕见的良性肿瘤（0.2%-1.6%）
- ◆ 肾脏混合性上皮间质肿瘤和成人性囊性肾瘤具有相似的年龄特征、重叠的组织学特点和免疫表型特征及相似的基因表达谱，2016版WHO肾脏肿瘤分类将两者归纳为MEST家族

Renal cell tumours		Mesenchymal tumours occurring mainly in adults	
Clear cell renal cell carcinoma	8310/3	Leiomyosarcoma	8890/3
Multilocular cystic renal neoplasm of low malignant potential	8316/1*	Angiosarcoma	9120/3
Papillary renal cell carcinoma	8260/3	Rhabdomyosarcoma	8900/3
Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma	8311/3*	Osteosarcoma	9180/3
Chromophobe renal cell carcinoma	8317/3	Synovial sarcoma	9040/3
Collecting duct carcinoma	8319/3	Ewing sarcoma	9364/3
Renal medullary carcinoma	8510/3*	Angiomyolipoma	8860/0
MiT family translocation renal cell carcinomas	8311/3*	Epithelioid angiomyolipoma	8860/1*
Succinate dehydrogenase-deficient renal carcinoma	8311/3	Leiomyoma	8890/0
Mucinous tubular and spindle cell carcinoma	8480/3*	Haemangioma	9120/0
Tubulocystic renal cell carcinoma	8316/3*	Lymphangioma	9170/0
Acquired cystic disease-associated renal cell carcinoma	8316/3	Haemangioblastoma	9161/1
Clear cell papillary renal cell carcinoma	8323/1	Juxtaglomerular cell tumour	8361/0
Renal cell carcinoma, unclassified	8312/3	Renomedullary interstitial cell tumour	8966/0
Papillary adenoma	8260/0	Schwannoma	9560/0
Oncocytoma	8290/0	Solitary fibrous tumour	8815/1
Metanephric tumours		Mixed epithelial and stromal tumour family	
Metanephric adenoma	8325/0	Cystic nephroma	8959/0
Metanephric adenofibroma	9013/0	Mixed epithelial and stromal tumour	8959/0
Metanephric stromal tumour	8935/1	Neuroendocrine tumours	
Nephroblastic and cystic tumours occurring mainly in children		Well-differentiated neuroendocrine tumour	8240/3
Nephrogenic rests		Large cell neuroendocrine carcinoma	8013/3
Nephroblastoma	8960/3	Small cell neuroendocrine carcinoma	8041/3
Cystic partially differentiated nephroblastoma	8959/1	Phaeochromocytoma	8700/0
Paediatric cystic nephroma	8959/0	Miscellaneous tumours	
Mesenchymal tumours		Renal haematopoietic neoplasms	
Mesenchymal tumours occurring mainly in children		Germ cell tumours	
Clear cell sarcoma	8964/3	Metastatic tumours	
Rhabdoid tumour	8963/3	The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) [917A]. Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours. The classification is modified from the previous WHO classification [756A], taking into account changes in our understanding of these lesions. *New code approved by the IARC/WHO Committee for ICD-O.	
Congenital mesoblastic nephroma	8960/1		
Ossifying renal tumour of infancy	8967/0		



01 研究背景

- ◆ MEST家族多表现为囊性病变，影像特异性低，与囊性肾癌难鉴别，部分病例存在过度诊治现象（如根治性肾脏切除）
- ◆ MEST家族术前定性诊断困难
- ◆ 肾脏穿刺活检有创，对囊性病变定性诊断价值有限（灵敏度和特异度均较低，33%-83%不等），具有易出血、假阴性、种植转移等风险
- ◆ MRI检查无创，软组织分辨率高，多平面、多序列、多参数成像
- ◆ Bosniak分类系统预测肾脏囊性病变良恶性，2019版基于MRI的Bosniak分类系统具有较高的诊断效能（83%）和良好的阅片者间一致性（ $Kw=0.68$ ）



02/ 研究目的及意义

研究目的

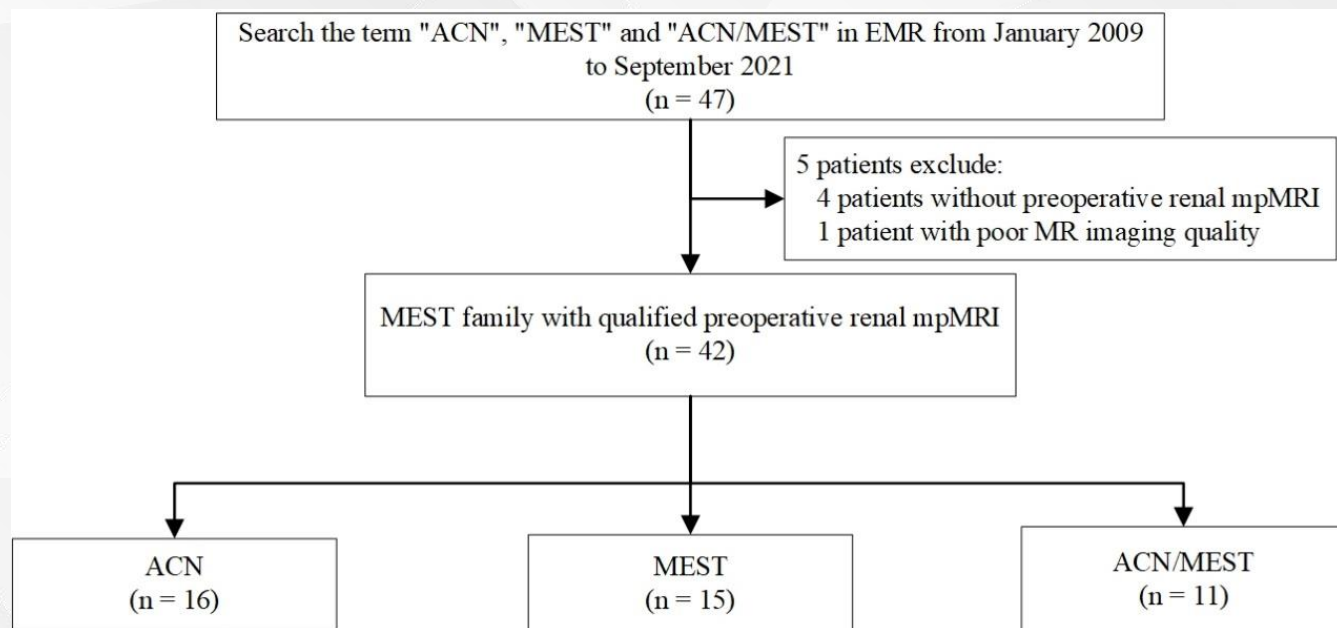
- ◆ 分析MEST家族的临床及MR影像特点
- ◆ 比较新旧版本Bosniak分类系统对囊性MEST家族的诊断价值

研究意义

- ◆ 提高MEST家族术前诊断准确性

03 材料与amp;方法-患者入组流程图

2名影像医师回顾性收集MEST家族患者临床、影像及病理资料



Flowchart showing the study selection process

ACN: adult cystic nephroma;

MEST: mixed epithelial and stromal tumor;

EMR: electronic medical record

纳入标准:

- ◆ 病理结果为MEST家族且病理结果完整
- ◆ 术前3个月内于本院行肾脏mpMRI检查
- ◆ 肾脏mpMRI序列完整 (T2WI、T1WI、DWI、多期动态增强及减影序列)

排除标准:

- ◆ 穿刺病理结果为MEST家族
- ◆ 图像质量不满足诊断需求 (如伪影明显、扫描野不全)



03/ 材料与amp;方法-MRI特征提取

- ◆ 两名泌尿生殖方向影像医师（2年和3年工作经验）独立评估记录MEST家族MR影像
- ◆ 评估内容：侧别、病变位置、病变最大径、病变形状、边界、生长方式、是否突向肾盂、是否具有假包膜、出血、强化特点等

临床	影像
<ul style="list-style-type: none">• 性别• 年龄• 主诉• 血尿史• 既往肿瘤病史• BMI•	<ul style="list-style-type: none">• 病变最大径• 形态• 生长方式• 假包膜• 出血• 囊壁强化特点 (结合减影图像)• 分隔强化特点 (结合减影图像)• 强化峰值•



03/ 材料与amp;方法-Bosniak分类评估

- ◆ 两名医师分别应用2005版和2019版 Bosniak 分类系统评估36例囊性 MEST家族，正式评估前均接受正规、系统的评估培训
- ◆ 评估结果不同时，协商达成一致

注：基于CT的2005版分类系统由本院两名高年资医生修改转化为基于MRI的版本

Category	Version 2005 of Bosniak classification system	Version 2019 of Bosniak classification system
I	Hairline-thin wall; signal intensity similar to cerebrospinal fluid; no sept or solid components; nonenhancing	Well-defined, thin ($\leq 2\text{mm}$) smooth wall; homogeneous simple fluid (signal intensity similar to cerebrospinal fluid); no septa or calcifications; the wall may enhance Three types, all well-defined with thin ($\leq 2\text{mm}$) smooth walls: <ol style="list-style-type: none"> Cystic masses with thin ($\leq 2\text{mm}$) and few (1-3) enhancing septa; any nonenhancing septa; may have calcification of any type Homogeneous masses markedly hyperintense at T2-weighted imaging (similar to cerebrospinal fluid) at noncontrast MRI Homogeneous masses markedly hyperintense at T1-weighted imaging (approximately $\times 2.5$ normal parenchymal signal intensity) at noncontrast MRI
II	Hairline-thin wall; few hairline-thin septa with or without perceived (not measurable) enhancement	Two types: <ol style="list-style-type: none"> Cystic masses with a smooth minimally thickened (3mm) enhancing wall, or smooth minimal thickening (3mm) of one or more enhancing septa, or many (≥ 4) smooth thin ($\leq 2\text{mm}$) enhancing septa Cystic masses that are heterogeneously hyperintense at unenhanced fat-saturated T1-weighted imaging
IIIF	Multiple hairline-thin septa or minimal smooth thickening of their walls or septa with or without perceived (not measurable) enhancement	One or more enhancing thick ($\geq 4\text{mm}$ width) or enhancing irregular (displaying $\leq 3\text{mm}$ obtusely margined convex protrusion[s]) walls or septa
III	Thickened or irregular wall or septa with measurable enhancement	One or more enhancing nodule(s) ($\geq 4\text{mm}$ convex protrusion with obtuse margins, or a convex protrusion of any size that has acute margins)
IV	Soft-tissue components (ie, nodule[s]) with measurable enhancement	



03/ 材料与amp;方法-统计与分析

- ◆ 应用SPSS 26.0和Stata/MP 16.0软件计算两版分类系统中的阅片者间加权Kappa值(Kw)
- ◆ Bosniak I-IV类分为低级别组（I、II和IIF）和高级别组（III和IV）
- ◆ Fisher精确检验分析比较囊性MEST家族在两版分类系统中分布是否具有差异
- ◆ $p < 0.05$ 差异具有统计学意义

注：Kw: ≤ 0.2 ，一致性较低
Kw: 0.21-0.4，一致性一般
Kw: 0.41-0.6，中等程度一致
Kw: 0.61-0.8，高度一致
Kw: 0.81-1.0，几乎完全一致



04 结果与讨论-患者人口统计学特点

The demographic features of MEST family

Variables	x ± S/n (%)
Age (Interquartile range)	40.9 ± 13.0
Gender	
Male	25 (59.5)
Female	17 (40.5)
Clinical presentation	
Incidental	33 (78.6)
Pain	9 (21.4)
RNS	
4 - 6	3 (7.1)
7 - 9	25 (59.5)
10 - 12	14 (33.4)
Surgery	
RN	9 (21.4)
PN	31 (73.8)
Cyst decapitating decompression operation	2 (4.8)
Recurrence/ Metastasis	
Yes	0 (0)
No	42 (100)

既往研究

- ◆ MEST家族好发于围绝经期女性（约46岁）
- ◆ 男:女=1:11 - 1:6

本研究

- ◆ MEST家族好发于中年患者
- ◆ 男:女≈1.5:1，无明显性别偏倚
- ◆ MEST家族多无临床症状
- ◆ 超过90%的MEST家族手术难度表现为中至高度复杂
- ◆ MEST家族均无复发和转移



04 结果与讨论-患者MRI特点

	x ± S/n (%)
Texture	
Cystic MEST family	36 (85.7)
Solid MEST family	6 (14.3)
Mean diameter (mm)	45.8 ± 22.4
Laterality	
Left	19 (45.2)
Right	23 (54.8)
Location	
Upper pole	8 (19.0)
Middle portion	27 (62.3)
Lower pole	7 (16.7)
Margin	
Well-circumscribed	31 (73.8)
Unclear	11 (26.2)
Shape	
Regular	13 (31.0)
Irregular	29 (69.0)
Growth pattern	
Exophytic	32 (76.2)
Endophytic	10 (23.8)
Renal sinus protruding	
Yes	20 (47.6)
No	22 (52.4)
Hemorrhage	
Yes	24 (57.1)
No	18 (42.8)
Pseudocapsule	
Yes	9 (21.4)
No	33 (78.6)

MRI characteristics of the MEST family

- ◆ 共纳入42例MEST家族（36例囊性，6例实性）
- ◆ 将近70%MEST家族形状不规则
- ◆ MEST家族多表现为外生性生长
- ◆ 约50%病例突向肾盂生长
- ◆ MEST家族不易出现假包膜



04/ 结果与讨论-MEST家族MRI强化特点

The enhancement pattern of the MEST family

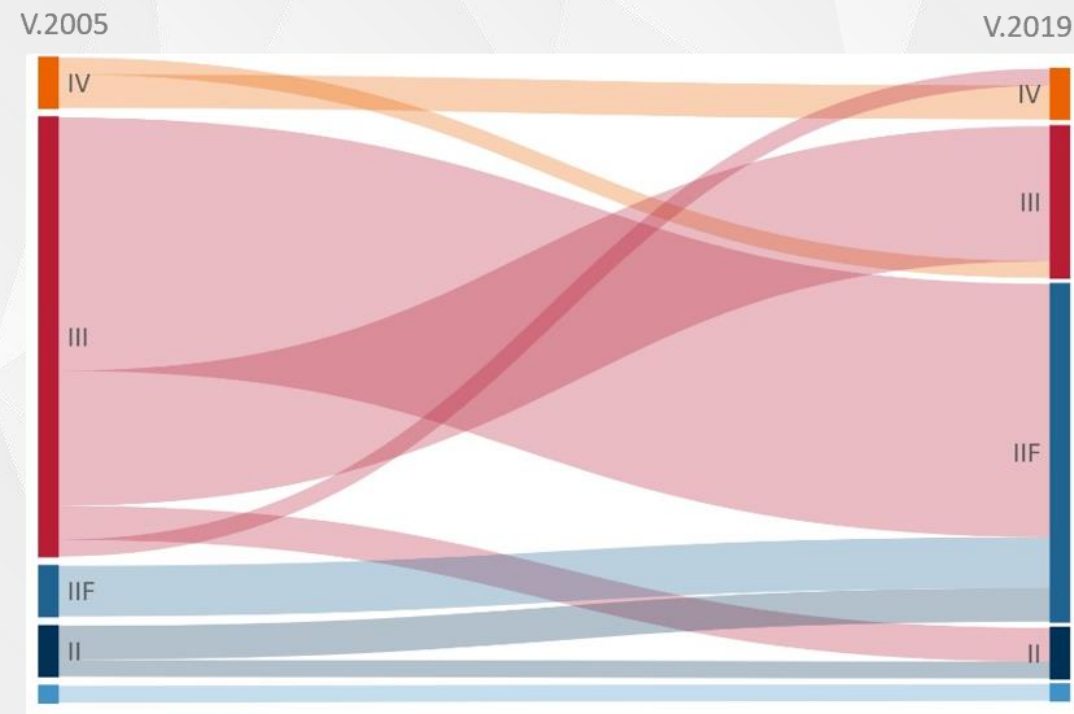
	Early enhancement	Delayed enhancement
Cystic MEST family		
Septa/Nodule	6	30
Wall	2	34
Solid MEST family	1	5

- ◆ 无论是实性还是囊性MEST家族，85%的病例为延迟强化
- ◆ 与典型的肾透明细胞癌的“快进快出”强化特点可鉴别

04 结果与讨论-MEST家族阅片者间一致性

Distribution of MEST family with Bosniak classification system, versions 2005 and 2019

	Low-grade			High-grade		<i>p</i> value
	I	II	IIF	III	IV	
BC system						< .001
Version 2005	1	3	3	26	3	
Version 2019	1	2	21	9	3	



- ◆ 相较于2005版Bosniak分类系统，2019版将更多囊性MEST家族分类到低级别（尤其是II和IIF类）
- ◆ 与2019版Bosniak分类系统初衷相符，将更多良性病变划分为低级别



04 结果与讨论-阅片者间一致性

Interobserver agreement between versions 2005 and 2019 of Bosniak classification system

		I	II	III	IV	k_w (95% CI)
Version 2005	Radiologists t 1	1	3	3	26	0.4221 (0.1587, 0.6674)
	Radiologists t 2	1	2	7	24	
Version 2019	Radiologists t 1	0	3	25	5	0.5026 (0.2330, 0.7721)
	Radiologists t 2	1	4	21	9	
<i>p</i> value						0.539

- ◆ 2019版Bosniak分类系统阅片者间一致性数值上高于2005版，但差异不具有统计学意义
- ◆ 可能与本研究样本量较小有关（36例囊性MEST家族）



MEST家族患病率无性别偏倚

2019版Bosniak分类系统将更多的囊性MEST家族划分到低级别

中年患者伴肾脏囊性病变，突向肾盂肾盂生长且延迟强化，应考虑MEST家族存在的可能性