

Supplemental Digital Content, containing 2 tables and 1 figure. The Supplemental Digital Content was not copyedited by *Archives of Pathology & Laboratory Medicine*.

Supplemental Table 1. Clinical data of 25 papillary fumarate hydratase (FH)-deficient renal cell carcinoma.

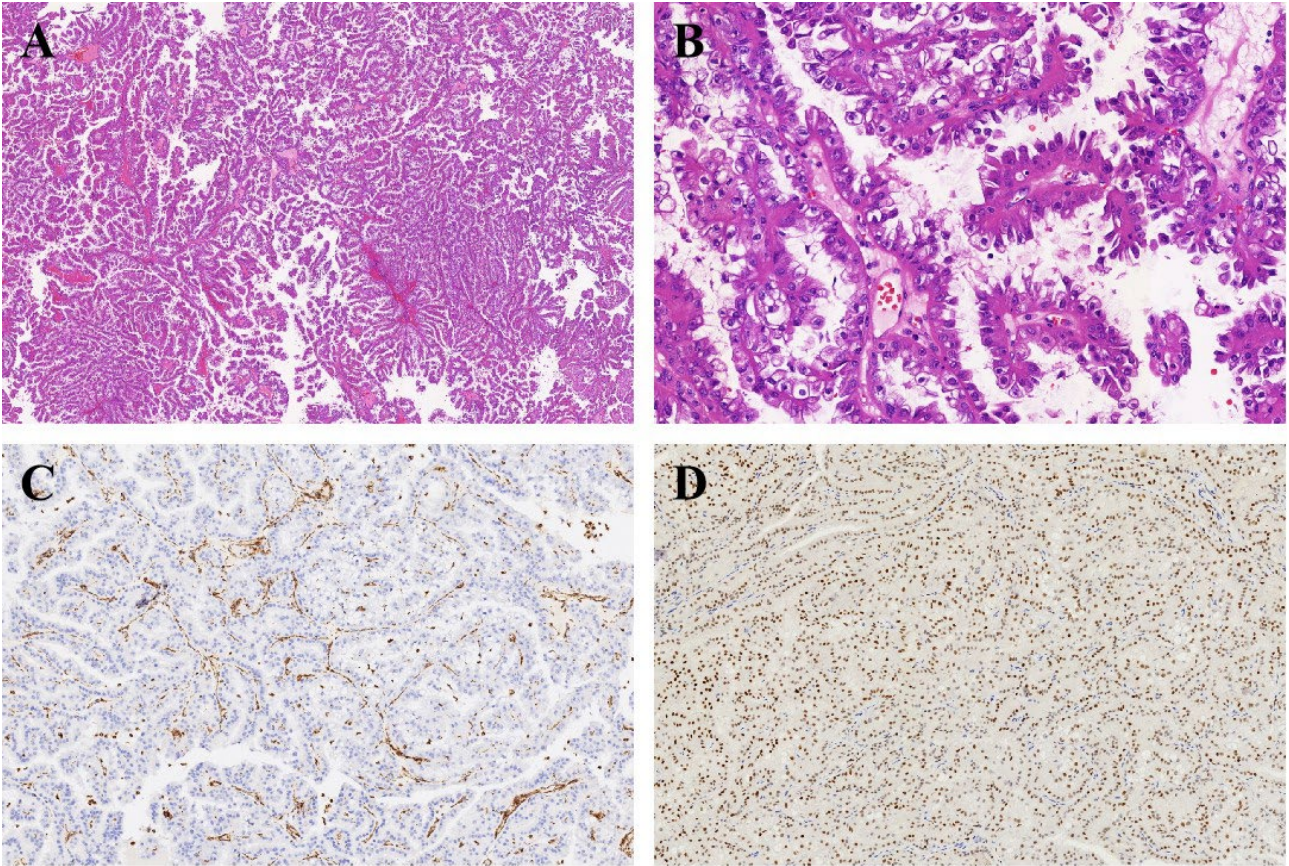
Case	Sex	Age/ year	Laterality	Size/ cm	Personal History of Leiomyomas	Follow-up / month	status
1	M	21	R	13	No	32	AWD
2	M	45	Bilateral	8	No	24	AWD
3	F	32	R	7.2	Uterine	38	AWD
4	F	35	R	4.5	No	25	DOD
5	M	34	Bilateral	5	No	13	AWD
6	M	64	L	3	No	16	AWD
7	F	32	L	10	Uterine	24	AWD
8	M	43	Bilateral	7.2	No	22	AWD
9	M	38	Bilateral	10.6	No	20	AWD
10	M	50	Bilateral	4.5	No	50	AWD
11	F	33	L	14	Uterine	24	AWD
12	F	26	R	7.7	/	20	AWD
13	M	53	L	5	No	12	DOD
14	M	41	R	4	No	66	DOD
15	M	53	L	4	No	29	DOD
16	M	54	Bilateral	8	Cutaneous	76	AWD
17	F	20	R	7	Uterine	52	AWD
18	M	30	L	7	No	17	AWD
19	M	29	L	6	No	41	AWD
20	F	53	L	8.6	Uterine	11	DOD
21	M	36	L	7	No	47	AWD
22	M	40	R	8	No	30	DOD
23	F	36	L	3.5	Uterine	60	AWD
24	M	22	R	8.6	No	56	AWD
25	M	34	L	11	/	23	DOD

AWD, alive with disease; DOD, died of the disease; M, male; F, female; L, left; R, right; /, not clear.

Supplemental Table 2. Integrated clinicopathological data of fumarate hydratase (FH)-deficient renal cell carcinoma (RCC) with predominant tubulocystic (n=10) or papillary (n=25) architecture.

		Tubulocystic FH-deficient RCC	Papillary FH-deficient RCC
age (year)		51.8±10.4	38.2±11.4
size (cm)		7.7±3.3	7.2±2.9
sex	male	8	17
	female	2	8
outcome	NED	2	0
	AWD/DOD	8	25
pure pattern	yes	0	2
	no	10	23
CK7	positive	2	4
	negative	8	21
FH	positive	0	2
	negative	10	23
2SC	positive	10	25
	negative	0	0
Ki67	<5%	0	0
	≥5%	7	17

FH, fumarate hydratase; RCC, renal cell carcinoma; AWD, alive with disease; DOD, died of the disease; NED, no evidence of disease.



Supplemental figure 1. Fumarate hydratase (FH) deficient renal cell carcinoma with pure papillary architecture. Tumor cells with eosinophilic cytoplasm and prominent nucleoli exhibited pure papillary architecture (A and B). FH was lost in the tumor cells (C) and GATA3 was diffusely positive in FH-deficient RCC (D) (hematoxylin-eosin, original magnifications $\times 40$ [A] and $\times 200$ [B]; FH, original magnification $\times 100$ [C]; GATA3, original magnification $\times 100$ [D]).