Case 4: 7-year-old boy

[Chief complaint]
Gross Hematuria

[History of present illness]
He visited an urological clinic because of gross hematuria from one day before the consultation. A renal tumor with intratumoral hemorrhage in the left kidney was suspected, and he was referred to our hospital for further examination and treatment.

[Past medical history]
Not particular
[Laboratory data]
WBC 6.2×10³/μl, RBC 4.48×10⁶/μl, HGB 11.9mg/dl ↓, PLT 22.2×10⁴/μl, TP 6.8g/dl, Alb 3.8g/dl ↓, T.bil 0.6mg/dl, ChE 305U/L, AST 27U/l, ALT 11U/l, LD 270U/l ↑, ALP 399U/l ↑, Na 138mmol/L ↓, K 3.9mmol/l, CL 102mmol/l, BUN 9mg/dl, Creatinine 0.33mg/dl ↓, Ca 9.1mg/dl, CRP 4.51mg/dl ↑

[Urinalysis]
Uric blood +3, Uric protein ±, RBC 87.7/HPF, WBC 5.9/HPF

→He was suspected of urinary tract infection by the laboratory studies and urinalysis performed at the first visit in our hospital.
Image presentation

- CT
- MRI
- 99mTc-DMSA renal scintigraphy
The abdominal CT shows three multilocular cystic masses in the left kidney.

These cystic masses have enhancing septa and no solid components.

The content of the cyst is slightly higher attenuation than that of water in the unenhanced CT.
The enhanced CT shows slightly low attenuated area (→) in the renal parenchyma around these cystic masses.

The hydronephrosis is not evident in the left kidney.

However, a part of these cystic masses appear to communicate with the renal pelvis (→).
MRI shows three multilocular cystic masses with hyperintense on T2WI in the left kidney, which have no solid component.

The hyperintense cysts on unenhanced fat suppression T1-WI (→) are seen within these masses.
The hyperintense cysts on unenhanced fat suppression T1WI show hyperintensity on $b=1000$ s/mm$^2$ diffusion weighted image (→) and have lower ADC values than the hypointense cysts on unenhanced fat suppression T1-WI, which may indicate hemorrhage.
Renal scintigraphy shows enlarged left kidney and some defects corresponding to these masses.
[Summary of imaging findings]

◆ Abdominal CT and MRI shows three multilocular cystic masses with contrast-enhancement septa and hemorrhage in the left kidney.

◆ Renal scintigraphy shows enlarged left kidney and some defects corresponding to these masses.

◆ The enhanced CT and MRI show slightly low enhanced areas in the renal parenchyma which were located around these cystic masses.
The differential diagnosis was cystic tumors occurring mainly in children.

- Multilocular cystic nephroma (MLCN)
- Cystic Wilms tumor
- Cystic partially differentiated nephroblastoma (CPDN)
- Segmental multicystic dysplastic kidney (MCDK)
◆ A left nephrectomy was performed with the pre-diagnosis of multilocular cystic nephroma.

◆ Gross specimen shows well-circumscribed tumor with multiple cysts (→).
Histopathological examination shows multiple noncommunicating cysts lined by flattened or cuboidal epithelium. These masses do not communicate with the renal pelvis.
The cyst epithelial cells are positive for CKae1/ae3 but negative for ER and CD10.
The final histopathological diagnosis

Multilocular cystic nephroma (MLCN)

Pediatric cystic nephroma

-WHO classification 2016: tumors of the kidney
WHO classification of tumors of the kidney

◆ Nephroblastic and cystic tumors occurring mainly in children
  - Nephrogenic rests
  - Nephroblastoma
  - CPDN

Pediatric cystic nephroma equivalent of MLCN

◆ Mixed epithelial and stromal tumor family
  - Adult cystic nephroma
  - Mixed epithelial and stromal tumor

※ Adult cystic nephroma is now thought to belong to the mixed epithelial and stromal tumor family and to be unrelated to pediatric cystic nephroma.
[Pediatric cystic nephroma]

◆ Definition:
- Multilocular, exclusively cystic neoplasm of very young children.
- The septa contain only fibrous tissue and differentiated tubules.

◆ Epidemiology:
- Occurrence more commonly in boys than in girls.
- Most patients are younger than 24 months old.
- No specific symptoms (flank pain, urinary infection, hematuria).
- Association with DICER1 mutations.

◆ Radiological features:
- Large multilocular cystic mass with varying size cysts, thin septa and no solid components.
- Cysts are non communicating with each other.
- Calcification and hemorrhage are rarely observed.
The differential diagnosis of pediatric nephroma

- CPDN
- Cystic wilms tumor

- The presence of solid components and thick septa supports the diagnosis of CPDN or wilms tumor. However, clearly distinction between pediatric cystic nephroma and these tumor is difficult.

- The presence of immature nephroblastic elements excludes diagnosis of pediatric cystic nephroma and indicates CPDN or cystic wilms tumor in the histopathology examination.
Correct answer

Multilocular cystic nephroma (MLCN)

Pediatric cystic nephroma

- WHO classification 2016: tumors of the kidney
References