



A rare cause of chronic tubulointerstitial nephritis in childhood: Questions

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Case

A 6-year-old boy was referred due to abnormal kidney function tests performed following vomiting and diarrhea lasting for 5 days before admission. The family denied a decrease in urine. There was no consanguinity between the parents nor did he have a known disease in his medical history. In his medical records, his serum creatinine was 0.6 mg/dL 6 months prior to admission. He had a body weight of 21.4 kg (25th–50th percentile), height of 118.5 cm (25th–50th percentile), body temperature of 36.7 °C, blood pressure of 158/100 mmHg (> 95th percentile +12), heart rate of 104/min, and respiratory rate of 26/min. The patient was oriented and cooperative. In his physical examination, he had +1 pretibial pitting edema. Respiratory sounds were normal. There was no splenomegaly or hepatomegaly. Laboratory tests are shown in Table 1.

A urinary catheter was inserted, but no urine was observed. The urine output was 0.3 cc/kg/h after forced diuresis. There was no finding in favor of active infection

in serology including COVID-19. Kidney biopsy revealed global sclerosis in 3; cellular/fibrocellular crescents in 4; fibrous crescents in 4; ischemic collapse in 10–11; and segmental sclerosis in 1 glomerulus of the 43 glomeruli. Basement membranes and mesangial cellularity were normal. A mixed type of inflammatory cell infiltration with dense lymphoplasmacyte cells, containing mild-to-moderate eosinophils, was observed in the tubulointerstitial area. In addition to active tubulitis findings including loss of brush border epithelium, localized shedding of epithelial cells, tubular dilatation with proteinous hyaline casts, moderate tubular atrophy and severe interstitial fibrosis attributable to chronic tubulointerstitial nephritis (CTIN) were noted (Fig. 1a and b). In the immunofluorescence study, there was +2 fine granular IgG staining in the basement membranes, and a segmental +1 non-specific IgM staining in the basement membranes in some glomeruli. Ophthalmologic examination was normal in the patient.

Questions

1. What could be the cause of chronic tubulointerstitial nephritis in this patient?
2. Which additional tests would you perform for the diagnosis?
3. What is the most likely diagnosis in this patient?

The answers to these questions can be found at <https://doi.org/10.1007/s00467-021-05326-y>

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Fig. 1 Diffuse interstitial inflammation including a mixed type of inflammatory cells (white arrows) tubular casts (asterisks), severe tubular atrophy and interstitial fibrosis (black arrows), with immature glomeruli (empty black arrow) (a H&E, original magnification $\times 40$) and a cellular crescent with periglomerular fibrosis (empty white arrow) (b H&E, original magnification $\times 100$)

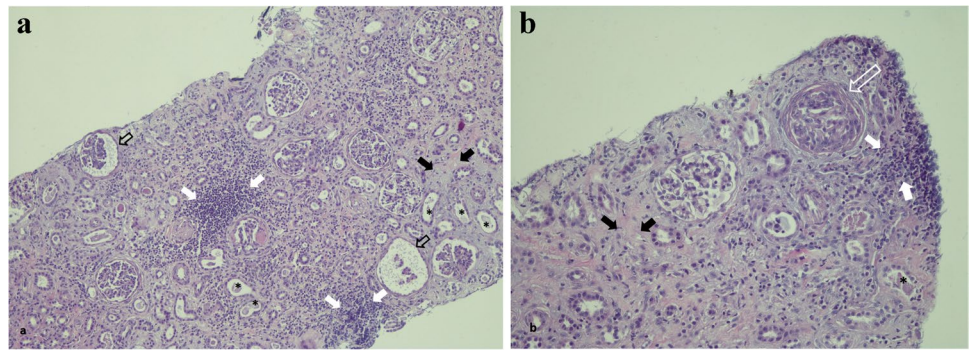


Table 1 Laboratory values of the patient at the admission

White blood cell count	$8.8 \times 10^3/\mu\text{L}$
Hemoglobin	12.4 g/dL
Platelets	$356 \times 10^3/\mu\text{L}$
Serum urea	198 mg/dL (N: 10–38)
Creatinine	6.2 mg/dL (N: 0.5–1.2)
Uric acid	6.4 mg/dL (N: 2–5.5)
Albumin	2.3 g/dL (N: 3.5–5.5)
Sodium	130 mmol/L (N: 134–150)
Potassium	4.91 mmol/L (N: 3.5–5.5)
Calcium	7.7 mg/dL (N: 8.8–10.8)
Phosphorus	6.6 mg/dL (N: 4–7)
Erythrocyte sedimentation rate (ESR)	108 mm/h (N: 0–20)
Parathormone	248 ng/L (N: 18.5–88)
C-reactive protein	4.6 mg/L (N: 0–5)
Blood gas	pH 7.13, pCO ₂ 48 mmHg, HCO ₃ 11.6 mmol/L, and BE – 16.7 mmol/L
Serum C3	1.23 g/L (N: 0.9–1.8)
Serum C4	0.3 g/L (N: 0.1–0.4)
IgG	8.44 g/L (N: 7–16)
IgA	1.75 g/L (N: 0.7–4)
IgM	0.76 g/L (N: 0.4–2.3)
IgE	24,600 IU/mL (N: 0–90)
Rheumatoid factor	Negative
ANA/anti-ds DNA	Negative/negative
Extractable nuclear antigen (ENA) panel	Negative
c-ANCA/p-ANCA	Negative/negative
Anti-glomerular basement membrane antibody	Negative
Phospholipase-A2 receptor antibody	Negative
Urine analysis	Specific gravity 1018; glucose +2; protein + 3; erythrocytes +2
Spot urine protein/creatinine	24.4
Tubular tests	
FeNa	3.1%
FeK	172%
TPR	45.6%

Declarations

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Conflict of interest

The authors declare no competing interests.