Pediatric Nephrology
https://doi.org/10.1007/s00467-022-05638-7

CLINICAL QUIZ

A patient presenting with fever after graft loss: Questions

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Received: 11 May 2022 / Accepted: 23 May 2022
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Keywords Fever · Pain · Kidney graft · Inflammation · Adolescent

Case report

A 15-year-old girl with kidney failure and on peritoneal dialysis presented with right flank pain and fever. Past medical history revealed that she was diagnosed with hypoplastic dysplastic kidney in the neonatal period. She underwent a living-related donor transplant with her father's kidney at 6 years of age. At 9th month post-transplant, mixed-type rejection developed and pulse methylprednisolone, antithymocyte globulin, intravenous immunoglobulin, and rituximab were administered. At 30th month post-transplant, hypodense mass lesions in the right and left hepatic lobes, the largest of which was 3.5 × 3.5 cm, were detected by routine ultrasonography screening; and histopathological examination was compatible with leiomyosarcoma. Then, immunosuppressive treatments were discontinued and chemotherapy was started. She lost her graft at the age of 13, prednisolone was continued at maintenance doses (5 mg/day), and peritoneal dialysis (PD) treatment was started. Twelve months later, she presented with a fever lasting 3 days and acute phase reactant elevation (Table 1). COVID PCR was negative; COVID Ig M and Ig G were positive. The patient was diagnosed with multisystem inflammatory syndrome in children (MIS-C) based on the *Centres for Disease Control and Prevention* criteria [1]. The patient's symptoms were as follows: fever, lymphopenia, laboratory evidence of inflammation, presence of pericardial effusion, and positive COVID serology. With the diagnosis of MIS-C, she received prednisolone (2 mg/kg/day), intravenous immunoglobulin, and low molecular weight heparin. As she was resistant to these therapies, an interleukin-1 receptor antagonist (anakinra) was given for 4 weeks.

Four months after the diagnosis of MIS-C, when she was receiving 20 mg/day prednisolone, she presented with fever and pain over the kidney graft. Her height was 141 cm (< 3 percentile), weight was 41 kg (< 3 percentile), and blood pressure was 130/65 mmHg. Physical examination revealed tenderness on the graft in the right pelvic area. The laboratory results at the diagnosis of MIS-C, after MIS-C, and at the admission with fever and pain are presented in Table 1. There were no leukocytes on microscopic examination of the peritoneal fluid, and no microbial growth was detected in dialysis effluent culture and urine culture. The nasopharyngeal swab real-time reverse transcription PCR for SARS-CoV-2 was negative. The patient was screened for common viral agents and mycobacterium tuberculosis, both of which were negative. On ultrasonographic examination, the size of the transplanted kidney, located at the right lower quadrant of the abdomen, was 120 mm; increased parenchymal echogenicity (grade I–II) was seen; no hydronephrosis was observed. Abdominal tomography did not reveal any finding supporting recurrence of malignancy.

Questions

1. What is your preliminary diagnosis for this patient?
2. Which tests will support the diagnosis?
3. What are the treatment options?

The answers to these questions can be found at http://dx.doi.org/10.1007/s00467-022-05646-7

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Published online: 14 June 2022

Table 1 Laboratory values of the patient

Variable	At the time of MIS-C diagnosis	After MIS-C treatment was completed	At the time of admission with fever and pain	Referrence range
Blood urea nitrogen, mg/dL	37	41	57	9–23
Serum creatinine, mg/dL	4.35	3.54	6.19	0.7–1.2
Serum albumin, g/dL	3.02	2.81	2.92	3.2–4.8
Hemoglobin, g/dL	8.5	12.3	9.4	12–16
Leukocyte count, mm^3	12,320	12,200	5890	4200–9500
Lymphocyte count, mm^3	1580	2080	1520	1300–3500
Platelet count, mm^3	312,000	227,000	199,000	150,000–450,000
C-reactive protein, mg/L	149	10.6	169	0–5
Erythrocyte sedimentation rate, mm/h	116	36	71	0–20
Procalcitonin, µg/L	1.12	0.69	0.49	0–0.05
Ferritin, µg/L	3980	1572	4550	13–150
Urine analysis	8.5	8.5	8.5	5–8
-pH	1015	1010	1007	1005–1030
-Density	+2	+1	+2	Neg
-Protein	4/hpf	2/hpf	4/hpf	<5/hpf
-Leukocyte				

MIS-C, multisystem ınflammatory syndrome in children

Declarations

Conflict of interest The authors declare no competing interests.

Reference

1. Centers for Disease Control and Prevention. Multisystem inflammatory syndrome in children (MIS-C) associated with coronavirus disease 2019 (COVID-19). https://emergency.cdc.gov/han/2020/han00432.asp

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