Lymphadenopathy, splenomegaly, intermittent neutropenia, and acute kidney injury: Questions

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Received: 25 June 2019 / Revised: 11 July 2019 / Accepted: 23 July 2019 / Published online: 29 August 2019
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Keywords Child · Anemia · Neutropenia · Splenomegaly · Lymphadenopathy · AKI · Hypertension

A 3-year-old female with an 18-month-old history of lymphadenopathy, intermittent neutropenia, and splenomegaly, presented to the emergency department with tachypnea and abdominal distention. Two weeks prior, she had experienced a bilateral purpuric rash, which resolved within a day, joint swelling, and non-specific abdominal pain. Blood work showed anemia (Hb 73 g/L) and neutropenia ($1.22 \times 10^9/L$), as well as acute kidney injury, urea at 15.2 mmol/L, and creatinine (Cr) at 34 $\mu\text{mol/L}$ (baseline Cr 18 $\mu\text{mol/L}$). There was no clear evidence of a hemolytic anemia, with a normal haptoglobin of 1.54 g/L, normal LDH of 724 U/L, and no schistocytes on blood film. Platelet count at presentation was normal at $217 \times 10^9/L$ and remained normal. Her serum albumin was a lower limit of normal at 35 g/L. Potassium was elevated at 5.7 mmol/L, magnesium at 1.2 mmol/L, and urate at 423 $\mu\text{mol/L}$. Her creatine phosphokinase was normal at 39 U/L.

From an immune standpoint, her ferritin was elevated at 223.5 $\mu\text{g/L}$. C3 and C4 were normal at 1.18 g/L and 0.23 g/L, respectively. IgA was elevated at 2.6 g/L, as was her IgG at 21.4 g/L, but IgM was normal at 0.6 g/L. ANA was positive at 1:160; anti-dsDNA, ANCA, and anti-GBM were all negative. Of note, her vitamin B12 levels in the serum were very elevated at >4427 pmol/L (normal 218–1305 pmol/L).

Urinalysis was positive for blood, but on microscopy, only 1–2 red blood cells were seen, with 0–1 white cells and no red cell casts. There was also mild proteinuria, with a protein/creatinine ratio of 60 mg/mmol and albumin/creatinine ratio of 25.9 mg/mmol. On examination, the patient's blood pressure was elevated at 123/86, and her respiratory rate (RR) was also increased at 44. A chest x-ray showed bilateral pleural effusions. There was no rash or evidence of joint swelling at time of presentation. Ultrasound showed splenomegaly and enlarged lymph nodes in the right upper quadrant at 1.3 cm. Her kidneys initially showed normal echogenicity and were normal sizes, of 8.4 cm on the right and 9 cm on the left.

Over the course of her admission, the albumin/creatinine ratio increased to 96.6 mg/mmol, serum albumin dropped to a nadir of 29 g/L, and creatinine peaked at 60 $\mu\text{mol/L}$. Her microscopic hematuria remained persistent. Her hypertension also remained an issue, requiring treatment with furosemide and amlodipine. Due to these features, a renal biopsy was performed.

Questions

- 1) What is your differential diagnosis?
- 2) What is the likely diagnosis?
- 3) What would you expect to see on renal biopsy?
- 4) How would you manage the patient?

The answer to this question can be found at <https://doi.org/10.1007/s00467-019-04322-7>.

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Compliance with ethical standards

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