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CLINICAL QUIZ

New-onset bone marrow aplasia in a 15-year-old adolescent with pauci-immune crescentic glomerulonephritis: Questions

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Case summary

A 15-year-old male adolescent of Ecuadorian origin had been adopted by a Swiss family at the age of 2 years. His past medical history was unremarkable, with no history of frequent respiratory illnesses, joint pain or skin problems. He was referred because dark urine and high blood pressure had appeared following a mildly febrile respiratory illness that had been treated with azithromycin and paracetamol. On examination he was well, fully conscious, normally hydrated, with a body weight of 52.1 kg, a blood pressure of 140/95 mm Hg, a respiratory rate of 12/min, and an oxygen saturation value of 96 %. The remaining examination was unremarkable.

The diagnostic work-up disclosed glomerular hematuria, moderate proteinuria (protein/creatinine ratio 184 mg/mmol; normal: ≲20 mg/mmol), a rather severe renal functional impairment (creatinine 256 μmol/L, urea 14.6 mmol/L), a normal chest X-ray, and negative blood tests for antinuclear, antineutrophil cytoplasmic, and antiglomerular basement membrane autoantibodies and complement C3 and C4.

A renal biopsy disclosed large cellular crescents in 14 out of 26 glomeruli. Immunochemistry for IgG, IgA, IgM, complement C3, complement C4, and fibrin showed the presence of fibrin in the crescents, but no other immunoreactants were detected. Electron microscopy did not disclose changes of the glomerular basement membrane, deposits or specific morphological changes. Hence, the diagnosis of antineutrophil autoantibody-negative pauci-immune crescentic glomerulonephritis was made.

Renal function improved (creatinine≈130 μmol/L) on treatment with oral candesartan, an angiotensin antagonist, 16 mg daily, four intravenous pulses of methylprednisolone 500 mg each (followed by daily oral prednisone), and 750 mg of intravenous cyclophosphamide once a month. Three weeks after the fourth dose of cyclophosphamide, treatment with oral azathioprine, an inhibitor of purine synthesis, 100 mg daily, was initiated. The dosage of azathioprine was halved 2 weeks later because of a tendency toward a low white blood cell count (4.2×10^9/L). One week after halving the azathioprine dose, the patient was admitted in poor general condition with moderate fever (38.3 °C), cough, severe respiratory distress, and a poor oxygen saturation value of 85 %. The diagnostic work-up disclosed a positive nasal swab for influenza A virus subtype H1N1 and very severe pancytopenia: hemoglobin 80 g/L, platelets 18×10^9/L, and total white blood cells count 0.1×10^9/L. A bone aspirate smear demonstrated a profoundly hypocellular bone marrow with a decrease in all cellular elements.

The answers to these questions can be found at http://dx.doi.org/10.1007/s00467-014-2849-9.

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Questions

1. What is the differential diagnosis for bone marrow aplasia in this patient?
2. Why was this patient predisposed to bone marrow aplasia?

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