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# A diagnostically challenging case of atypical hemolytic uremic syndrome in the course of severe hypertension

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**Introduction:** Atypical hemolytic uremic syndrome (aHUS) is a rare, progressive, often genetic disease that can be fatal if untreated. It may manifest with proteinuria, hematuria without acute kidney injury (AKI). It frequently leads to refractory hypertension, AKI and myocardial infarction or encephalopathy.

We present a case of a 51-year-old patient, treated twice in the Clinic of Internal Medicine, Nephrology and Dialysis Therapy of the Military Institute of Medicine – National Research Institute, with refractory hypertension and AKI, who developed a-HUS, in the course of hypertensive nephropathy with renal failure.

**Case description:** A 51-year-old patient presented to the Emergency Department with a 1-month history of abdominal pain and intermittent vomiting for 5 months. On admission, the patient was in good general condition but in hypertensive crisis (blood pressure: 248/145 mmHg, heart rate: 121 bpm) requiring intravenous urapidil infusion. Severe renal failure was identified with a creatinine level of 5.7 mg/dl, urea 140 mg/dl and hypokalemia $K^+$ 2.2 mmol/l. Furthermore, normocytic anaemia, thrombocytopenia, proteinuria and hematuria were observed. A multidrug antihypertensive regimen was initiated. The patient developed atrial fibrillation during hospitalisation, managed with amiodarone; echocardiography showed features of left ventricular hypertrophy, and head computed tomography showed diffuse white matter hypodensity. Labs indicated secondary hyperparathyroidism. A right kidney biopsy was performed, complicated by retroperitoneal hematoma with subsequent hypotension and anemization, however without signs of active bleeding. Amoxicillin with clavulanic acid was included in addition to 3 units of packed red blood cells. After radiologic consultation, angiography with additional renal artery embolization. The 50% hemolytic complement (CH50) and a disintegrin-like and metalloprotease with thrombospondin type 1 motif 13 (ADAMTS13) activity levels are not without abnormalities. Genetic screening for aHUS was initiated. Renal biopsy histopathology revealed arterionephrosclerosis, pointing to a hypertensive aetiology. Intravenous iron and folic acid supplementation were started for the mixed-aetiology anaemia. A month later readmission was due to high renal parameters, and acidosis was identified, along with anaemia and thrombocytopenia. Low complement C3 level 77 mg/dl. Genetic testing confirmed aHUS. However, the patient was not qualified for ravulizumab therapy by the National Commission.

**Conclusions:** The aHUS is a diagnostically challenging disease due to non-specific symptoms and multi-organ manifestations. This case illustrates one of the mechanisms in which aHUS may develop in the course of severe hypertension, which, in the setting of aHUS, was particularly refractory to treatment.