O_14 DOI: https://doi.org/10.5114/reum/219191

Secondary Evans' syndrome as a first presentation of systemic lupus erythematosus: case study

Jakub Góra[1,2], Stanisław Niemczyk[3]

[1]Student Scientific Association of Nephrology at Clinic of Internal Medicine, Nephrology and Dialysis Therapy, Military Medical Institute – National Research Institute, Warsaw, Poland

[2]Medical Faculty, Medical University of Warsaw, Poland

[3]Clinic of Internal Medicine, Nephrology and Dialysis Therapy, Military Medical Institute – National Research Institute, Warsaw, Poland

Key words: Evans' syndrome, SLE, nephrotic syndrome, AIHA, ITP

Introduction: Evans' syndrome is a rare condition defined as concurrent autoimmune hemolytic anaemia (AIHA) and immune thrombocytopenia (ITP). Distinction has been made between primary and secondary variant, which differentiate it on bases if condition is idiopathic or associated with another systemic disease. The most common causes of secondary variant include: haematological malignancies and autoimmune disease, especially systemic lupus erythematosus (SLE). This condition could sometimes precede the onset of the underlying disease.

Case description: A patient aged 43 years was admitted to the nephrology ward in March 2025 with massive oedema and acute kidney injury. She was treated since 2001, the first symptom was thrombocytopenia. Diagnosis of ITP was made, the first line of treatment was vincristine, stopped due to polyneuropathy. Then she was managed with glucocorticosteroids (GCs), due to resistance, the splenectomy was performed, after which the level of platelets of 50–90 G/l was achieved. In 2014, she developed AIHA and a recurrence of thrombocytopenia. The antinuclear, anti-GPIa/II and anti-GPIIb/IIa antibodies were detected. In bone marrow biopsy, hypoplasia was described corresponding to an autoimmune process. Concurrent infection of parvowirus B19 was detected. The patient was treated with GCs, intravenous immune globulin and multiple blood transfusions. Between 2015 to 2020, patient experience few recurrences, all treated with GCs and blood transfusions. As complications to coagulation disorders, the patient underwent paralytic ileus in 2015, ischemic stroke in 2018 and cardiac infarction in 2019. During neurological examination in 2017, anti-AQP4 antibodies were detected, disease from neuromyelitis optica spectrum disorders was diagnosed. The GCs were administrated with a good response. During hospitalisation in nephrology, in laboratory finding proteinuria in the nephrotic range was detected and estimated glomerular filtration rate of 50 ml/min/1.73 m^2. Renal biopsy was impossible due to coagulation disorders. The chronic kidney disease on the basis of lupus nephritis, was diagnosed. The patient was treated according to the EuroLUPUS scheme with 500 mg of cyclophosphamide. After 6 cycles, partial remission of nephrotic syndrome was achieved.

Conclusions: Systemic lupus erythematosus is a heterogeneous disease. Solely haematological manifestation preceding other symptoms posed a diagnostic challenge in this case. Renal involvement and good response to EuroLUPUS treatment support the diagnosis of SLE. The most important issue in this patient is treating the underlying disease.