DIFFICULTIES IN DIAGNOSIS AND THERAPY MANAGEMENT OF EVANS SYNDROME

Mihaela Lebedenco, Nicoleta Berbec, Elena Coles, Diana Mandescu, Cristina Negotei, Anca Lupu
UMF Carol Davila, Bucharest, Romania;
Coltea Hospital, Bucharest,Romania

Background: Evans syndrome was first described in 1951 and defined as autoimmune disease characterized by simultaneous or sequential development of autoimmune hemolytic anemia and autoimmune thrombocytopenia with or without immune neutropenia in the absence of any known causes.

Material and Methods: Patient C.C. is known with Evans syndrome at the age of 30 years (1997), also associated with autoimmune thyroiditis at the age of 30 years (Euthyrox therapy). At first admission showed marked physical fatigue syndrome, bleeding (epistaxis, gingival bleeding, meno-metrorrhagia), apparently recently installed after intercurrent respiratory and clinical examination: obesity, pallor, scleral jaundice, bruising and generalized purpuric rash, haemoragic bullae on the mouth without peripheral lymphadenopathy without hepatosplenomegaly. In order to establish the correct diagnosis was made biological investigations, tests of hemolysis, cytogenetic and imaging.

Results and discussion: Positive diagnosis was Evans syndrome, which has been conducted over time corticosteroids, immunosuppression (Cyclophosphamide, Imuran), Vinca rosea alkaloids, splenectomy and thrombopoietin receptor agonists. Is interesting association of the Evans Syndrome with autoimmune thyroiditis, persistent normalization of Hb postsplenectomy with thrombocytopenia, hemolysis and brutal response compensation at the initiation with Revolade. Management of Evans syndrome treatment remains a challenge. Although we try a number of new molecules in the disease, corticosteroids remain the first-line choice and. It remains to prove the effectiveness and superiority of treatment with thrombopoietin receptor agonists in this autoimmune disease.