

Evans Syndrome Secondary to Sars-Cov2 Infection: Case Report

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Case History: In January 2022 a 90-year-old woman was admitted to our ward for severe pancytopenia especially as regards red blood cells (Hb: 3.7 g/dl) and platelets (7000 u/L). In the medical history the patient presented arterial hypertension well controlled by therapy with Ramipril 5 mg, type 2 diabetes mellitus treated with metformin 500 mg; moreover, the patient recently contracted Sars-CoV2 infection, which was negativized in December 2021. The physical examination showed: pale skin, no signs and symptoms of bleeding, normal vital parameters. At the entrance, the exams were almost normal except for the blood count and hypoproteinemia (5.5 g / dl). The patient underwent several transfusions of concentrated red blood cells as well as platelet pools as supportive therapy. On CT abdomen with contrast medium, the presence of multiple lymphadenomegalies in correspondence with the small gastric curvature was highlighted. The patient underwent, on the recommendation of the haematologist specialist, a bone medullary biopsy and a flow cytometric examination on peripheral blood, which did not show any significant changes. Finally, direct and indirect Coombs tests and anti-platelet antibodies were performed on peripheral blood, which showed positivity for direct polyspecific Coombs test, IgG species and anti-platelet antibodies. We therefore made a diagnosis of Evans Syndrome possibly secondary to previous Sars-CoV2 infection. At this point the patient was subjected to first-line therapy with prednisone at a therapeutic dosage of 1 mg per kg per day (75 mg). After only two days of full-dose steroid therapy, hemoglobin rose to 8.8 g / dl and platelets to 64,000 / uL. Therefore we decided to discharge the patient with the indication

to continue the steroid therapy at the aforementioned dosage and to recommend subsequent haematological outpatient checks.

Discussion and Conclusions: Evans syndrome is defined as the concomitant or sequential occurrence of immune thrombocytopenia (ITP) and autoimmune haemolytic anaemia (AIHA). The anaemia is an AIHA due to warm antibodies that are usually of IgG isotype, exceptionally IgA, thus excluding cold agglutinins. It's a rare disease with an annual incidence of 1.8/million person-years. AIHA is suspected in case of anaemia associated with reticulocytosis and with markers of haemolysis (elevated LDH, low haptoglobin and elevated indirect bilirubin). ITP remains a diagnosis of exclusion suspected in case of rapid onset thrombocytopenia not related to liver diseases, splenomegaly, drug-related thrombocytopenia, bone marrow deficiency or inherited thrombocytopenia. The diagnostic procedure of Evans 'syndrome must exclude differential diagnoses and determine the primary or secondary nature of Evans 'syndrome. Recently among the secondary etiologies of Evans 'syndrome has been reported SARS-CoV-2 infection. Corticosteroids represent the cornerstone therapy, used at daily dose of 1mg/kg of prednisone. Second-line therapy include: IVIg, rituximab, splenectomy, immunosuppressant and stem cell transplantation.

Currently the patient is in follow-up at the Hematology clinic of our company and at the first checks performed the levels of hemoglobin and platelets are constantly increasing; therefore she has started to scale down steroid therapy.