Abstract

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Auto-immune haemolitycc anemia (AHIA) and thrombocytopenia, Evans'syndrome?

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Background

Evans' syndrome is defined as the concomitant or sequential association of warm auto-immune haemolytic anemia (AIHA)and immune thrombocytopenia (ITP).Etiology is unkonwn,sometimes is associated to another disease, particularly with haematologic malignacies,primary immunodeficiencies, thrombotic microangiopathies, anaemia due to bleedings complicating ITP, myelodysplastic syndromes.

Case history

A 67- years old caucasian man presented to the Emergency Department reporting diffuse abdominal pain,fatigue, exertional dyspnea, fever, jaundice. On physical examination he presented with tachypnea, tachycardia, oxygen saturation 86% in ambient air, so he started O2 therapy with VMK 60% Fio2. Its right foot appeared pale and pulseless, so he immediately performedCT angiography oh thechest,abdominal and inferior limbs who revelealing multiple parenchimal consolidations bilaterally, obclusion of mesenteric artery, iliac and right tibial artery. Empirical antibiotical therapy,togetherCilostazolwas started0 His past medical history was significant for hypertension, polyvascular atherosclerotic disease, type II diabetes mellitus, chronic hepatopathy HCV-HBV related. At admission to the Internal Medicine ward the patient presented with fever, Glasgow coma Scale of 11 points (E 4 M4 V3), eupnoic in O2 therapy VMK 60%. Onphysical examination crackles on basal lungs, diffuse abdominal pain, pale,scleral jaundice. Laboratory exams showedHb 5.3 g/dL(therefore he received multiple blood transfusions, with an increase of haemoglobin to 7.5), leukocitosis, PCT 34 ng/ml,CPR 11 mg/dL. Diagnosis of sepsis (Qsofa 3)and thromboangiitis obliteranswere made, and iloprost, dual antiplatelet with Clopidogrel/Aspirin and fluid correctionwere started. Despite repeated blood trasfusion, hemoglobin concentration decreased with concomitant increase of LDH,bilirubin and reduced haptoglobin.Coombs'test was positive for IgG antibodies.Thus, AIHA diagnosis was made, but severe thrombocytopenia was found at haemochromocytometric examination, so Evan's syndrome was suspected.To rule out secondary causes of Evans' Syndrome, total body CT scan with contrast,serum protein electrophoresis, autoimmunity panel, screening for viral causes was performed,all with a negative result.No underlying pharmacological cause was found. Peripheral blood smears revealed lymphopenia, presence of immunoglobulins, some cells defined as "blasts/abnormal lymphocytes". Immunosuppressive therapy with high dose of corticosteroids was started, 1mg per kg for 4 week, then slow tapered over the following six months. Prophylaxis therapy with Aciclovir and Sulfamethoxazole/Trimetropim was started. After one week from therapy there was significant improvement in symptomatology and laboratory test. Haemoglobin was stable, so no more blood transusions were made. The cause of AHIA and thrombocytopenia remained uncertain, so the patient was eventually referred to a hematology department to perform a bone marrow biopsy.

Discussion

AIHA is suspected in case of anaemia associated with reticulocytosis and with markers of haemolysis(elevated LDH, low haptoglobin and elevated indirect bilirubin) with a positive direct antiglobulin test for IgG with or without complement (C3d) as cold agglutinins are excluded from ES. ITP remains a diagnosis of exclusion. In this clinical case, differential diagnosis with thrombotic microangiopathies is more important, but patients were treated with steroids, which did not improve cytopenia. In Evans' syndrome corticosteroids represent the cornerstone therapy, used at a daily dose of 1 mg/kg of prednisone. The duration of treatment is 3–4 weeks with slow tapering over six months.