SKIN MANIFESTATIONS OF INTERNAL DISEASE

CUTANEOUS THROMBOSIS ASSOCIATED WITH PAROXYSMAL NOCTURNAL HEMOGLOBINURIA

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Background: Paroxysmal Nocturnal Hemoglobinuria (PNH) is a chronic acquired clonal stem cell disorder manifesting as hemolysis, thrombosis and bone marrow failure. Cutaneous manifestations, due to thrombosis of dermal blood vessels, are uncommon and thromboembolic complications constitute the cause of morbimortality in PNH.

Observation: We report a case of a 35-year-old patient, with a previous history of bone marrow aplasia, which developed multiple purpuric plaques with central hemorrhagic blister and necrosis involving ears, face, trunk, upper and lower limbs. Laboratory analyses revealed hemolytic anemia (hemoglobin: 7.7 g/dl; reticulocytes: 7.2%; LDH: 1500 UI/l; haptoglobin: 93 mg/dl), leukopenia, thrombocytopenia, elevated D-dimer (5363 ng/ml) and hemoglobinuria. Cryoglobulins, antineutrophil cytoplasmic antibody, anticardiolipin immunoglobulin, extractable nuclear antigen, anti-hepatitis B virus antibody, anti-hepatitis C virus antibody and HIV antibody were negative. Skin biopsy showed fibrin thrombi in dermal blood vessels and extravasated red blood cells. Direct immunofluorescence revealed IgM, C3 and fibrin deposits. Bone marrow biopsy demonstrated hypocellular marrow. The diagnosis of PNH was confirmed by CD157 flow cytometry and FLAER with 78.3% of neutrophils and 61.4% of monocytes being abnormal. Corticosteroid and anticoagulation with Enoxaparin were initiated with a partial response until the administration of Eculizumab with which the improvement of the skin lesions was obtained.
Thrombotic events in PNH often affect unusual sites and skin manifestations occur in 0.6% of cases. It is a chronic illness but there may be periodic exacerbations precipitated by infection, surgery or drugs. Eculizumab, an anti-C5 monoclonal antibody, is the treatment of choice for patients with severe PNH, although allogeneic hematopoietic cell transplantation remains the only
curative therapy.

Key message: The clinical case of our patient is exposed due to the low prevalence of this disease of complex diagnostic and therapeutic approach, which demonstrated an excellent response to the therapy established.