Abstract

Health Science Journal
ISSN 1791-809X

2021
Vol.15 No.P13

Moschowitz Syndrome: Case Report

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Background: Moschowitz syndrome is an acute syndrome characterized by purpura, hemolytic anemia and thrombocytopenia. The incidence is 0,4/100000/year. In contrast of hemolytic-uremic syndrome, it shows neurological disease in 90% of the cases, and not always renal insufficiency. This syndrome is characterized by ADAMTS13 protein deficiency; so the ultralarge multimeter of the von-Willebrand factor, secreted by activated endothelial cells, is not appropriately cleaved between Tyr1605-Met1606, causing spontaneous and massive platelet aggregation in many organs, exept lung. It's caused by a genetic mutation of 9q34 gene or it's acquired for immune mediated mechanisms.

Case history: A 47 years old male presented anaemia (Hb: 6,6 g/dL), PLT 5000, headache, fever, confusion and petechiae. Blood workup showed total bilirubin 4,2 mg/dL, with indirect bilirubin 2,9 mg/dL, increased levels of LDH and ferritin, haptoglobin 8 mg/dL. ADAMTS13 activity assays depicted reduction of its activity (0,2%) with inhibitory levels (9,28 BU/mL). In three days diagnosis was made.

Discussion: The differential diagnosis includes SEU, CID, HELLP syndrome, HIT. The therapy is plasmapheresis; in refractory PTT Rituximab, an anti CD20 monoclonal antibody, and Caplacizumab, an inhibitor of interaction between von Willebrand factor and platelets. It's necessary an early diagnosis for its poor prognosis.