Afectarea multiorganica in microagiopatiile trombotice PTT-like – implicatia patogenica a microtrombozelor vasculare

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Microagiopatiile trombotice sunt afectiuni rar intalnite in practica clinica curenta, caracterizate prin ocluzii trombotice la nivelul microcirculatiei, consecinta acestora fiind aparitia trombocitopeniei de consum, a anemiei hemolitice microangiopatice, insotite de semne de afectare ischemica multiorganica. Daca forma clasica de purpura trombotica trombocitopenica (PTT), care presupune asocierea anemiei hemolitice autoimune si a trombocitopeniei, asociaza in mod frecvent afectare renala si neurologica, in sindroamele PTT –like exista manfestari hematologice similare, insa cu afectare multiorganica. Elementul comun al microagiopatiilor trombotice (PTT si PTT-like) este afectarea vasculara microtrombotica diseminata, secundara aparitiei de microtrombi formati din plachete si multimeri de dimensiuni mari de factor von Willebrand, insa mecanismul patogenic de aparitie al acestora este diferit, cu implicatii terapeutice diferite in functie de factorul etiologic.

Thrombotic microangiopathies are rare disorders characterized by thrombotic occlusion of the microcirculation that cause consumptive thrombocytopenia, microangiopathic haemolytic anaemia and signs of ischaemic damage in different organs. Thrombotic thrombocytopenic purpura (TTP) is characterized by microangiopathic hemolytic anemia and thrombocytopenia associated with brain and kidney dysfunction. TTP-like syndrome occurs with similar hematologic changes, but with multi organ dysfunction syndrome. The vascular disseminated microthrombotic disease is the key element for TTP and TTP-like syndrome, both are characterised by microthrombi composed of platelet-unusually large von Willebrand factor multimers complexes. The difference betweend these two syndromes is the underlying pathogenic mechanism leading to vascular microthrombotic disease, with diffrent treatment options.