

VISCOELASTIC TESTS (ROTEM) IN DISSEMINATED INTRAVASCULAR COAGULATION ASSOCIATED WITH AORTIC ANEURYSM.

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Background. Aortic aneurysms can be complicated by disseminated intravascular coagulation (DIC) with a predominantly fibrinolytic type in which both coagulation and fibrinolysis are simultaneously activated. The procoagulant state may contribute to microvascular and macrovascular thrombosis but in “enhanced-fibrinolytic-type DIC” multiple fibrin clots dissolve one after other due the hyperfibrinolysis. As a result, ischemic organ damage due to multiple microthrombosis is rarely seen as a clinical manifestation; in contrast, severe bleeding symptoms are more likely to occur with the dissolution of the thrombi. Characteristic laboratory findings include a low platelet (PTL) count, a normal-to-prolonged prothrombin time (PT) and a shortened-to-prolonged activated partial thromboplastin time (aPTT). Due to the enhanced fibrinolysis, the D-Dimer is increased (are especially high the Fibrinogen degradation products, FDP) and Fibrinogen levels are consequently decreased. In addition levels of Plasminogen Activator Inhibitor-1 (PAI-1 a fibrinolytic inhibitor) are normal or only mildly elevated. Concentrations of α 2plasmin-inhibitor (α 2PI) are markedly decreased and when they are less than 50%, caution should be taken against major bleeding. Fibrinogen levels are also markedly decreased in typical cases, not only because of the consumption associated with the dissolution of multiple microthrombi, but also because of the degradation of Fibrinogen by Plasmin.

Cases Report. We describe two cases of “enhanced-fibrinolytic-type-DIC” associated with aortic thrombosed aneurysms. Both patients presented with bleeding symptoms (spontaneous large limbs hematomas and anemia) associated with

marked reduction in Fibrinogen (< 100 mg/dl), prolonged PT/aPTT, mild thrombocytopenia and elevated D-dimer. The “enhanced-fibrinolytic-type-DIC” diagnosis was made even through viscoelastic tests (ROTEM). The table shows the clinical and laboratory findings of the patients. The cornerstone of DIC treatment is the treatment of the underlying pathology. However in both patients the surgery was not feasible. Therefore the therapeutic approach was to restore Fibrinogen and antifibrinolytic therapy in the acute phase of haemorrhage and antiplatelets/anticoagulant therapy once the bleeding is resolved.

Conclusions. It is known that some vascular diseases such as hemangiomas and vascular malformation (Kasabach-Merritt syndrome, Klippel-Trenaunay-Weber syndrome) can be associated with a chronic hyperfibrinolytic DIC and also in some aortic aneurysms, especially if large or complicated (with endoleak or thrombosis). In “enhanced-fibrinolytic-type DIC” associated with aortic aneurysms activation of coagulation factors and plasmin are responsible for both the state of hypercoagulability and fibrinolysis, with increased thrombotic or bleeding risk. This process can be dynamic and may require simultaneous haemostatic-replacement, but also anticoagulant or antiplatelet treatment in combination or later. The use of the ROTEM in both cases confirmed the hyperfibrinolytic state and allowed the correct administration of Fibrinogen and Antifibrinolytic therapy, proving to be a valid aid even outside the his codified use sets (polytrauma, post-partum hemorrhage, cardiac surgery and liver transplant).

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The table shows a description of the two cases compared.

	Case 1	Case 2
Age (Sex)	74 years (Male)	73 years (Male)
Medical history	<ul style="list-style-type: none"> No history of smoking Thyroidectomy for goiter without bleeding Psoriatic arthropathy of the right knee and quadriceps (treated with Methotrexate, Hydroxychloroquine and Adalimumab in 2023) Hypercholesterolemia Mild thrombocytopenia (70-80000/mmc) since 2015 (BGM negative). 2023 right inguinal empioplasty complicated by wall hematomas: PTL 55 000/mmc pre-surgery (pre-surgery prophylactic transfusion) 	<ul style="list-style-type: none"> Previous smoking Multiple Myeloma IgG/A undergoing autologous stem cell transplant in negative FU Hypothyroidism in replacement therapy Chronic ischemic heart disease revascularized in 2018 (PTCA/DES) in SAPT Protective colostomy in previous inguinal hernia surgery Chronic renal disease (G3b)
Type of Vascular Aneurysm	Fusiform abdominal aortic aneurysm with parietal thrombosis (4 cm of diameter)	2022 subclavian endoprosthesis for thoraco-abdominal aortic aneurysm with bulky residual endoleak at DG-D7 and further sub-renal aneurysm (both 6 cm of diameter)
Symptoms at onset	June 2024: spontaneous skin hematomas on the limbs, large in size and causing anemia.	September 2023: large subcutaneous hematoma right lower limb with severe anemia (Hb 4.3 g/dl). Previous epistaxis and gingival bleeding (stop antiplatelet therapy)
Laboratory tests	<p>Hb 10.6 g/dl PTL 37 000/mmc PT 1.29/aPTT 1.32 Fibrinogen 69 mg/dl - D-Dimer 22 891 mcgr/L LAC negative</p> <p>Coagulation factors assay</p> <ul style="list-style-type: none"> FVIII 98%, FvW Ag 161% FIX 110%, FXI 102%, FXII 74% FVII 73% FV 71%, FX 99% FII 110% FXIII 42% α-2 antiplasmin < 50% <p>Renal function normal Epatic function normal</p>	<p>Hb 4.3 g/dl PTL 95 000/mmc PT 1.19/aPTT 1.83 Fibrinogen 47 mg/dl - D-Dimer 31.23 mg/L LAC negative</p> <p>Coagulation factors assay</p> <ul style="list-style-type: none"> FVIII 82%, RICOF 1.79 U/ml FIX 147%, FXI 87%, FXII 85% FVII 133% FV 69%, FX 132% FII 89% FXIII 73.4% α-2 antiplasmin 130% <p>Renal function Crea 2 mg/dl (31 ml/min) Epatic function normal</p>
ROTEM		
Other Tests	Total Body TC negative PET negative BGM negative (complicated by late subcutaneous hematomas) Colonoscopy negative	Total Body TC negative Hematologic test (negative Bence Jones proteinuria, no monoclonal component)
Therapy	Fibrinogen 4 g ev Tranexamic acid ev and oral	Fibrinogen 4 g ev Tranexamic acid ev and oral Antiplatelet therapy 75 mg quickly discontinued due to mucosal bleeding
Complications	Superficial venous thrombosis of the right basilic vein at the catheter site treated with Enoxaparin 4000 U die	2024 spontaneous hemarthrosis right shoulder
Follow-up 1 month	PTL 73 000/mmc PT 1.17 e aPTT 1.05 Fibrinogen 403 mg/dl - D-Dimer 13 720 mcgr/L	PTL 103 000/mmc PT 1.23 e aPTT 1.58 Fibrinogen 89 mg/dl - D-Dimer 30 U/L