

Coagulazione intravascolare disseminata

A RARE CASE OF ACUTE PROMYELOCYTIC LEUKAEMIA PRESENTING WITH A PURELY THROMBOTIC DISSEMINATED INTRAVASCULAR COAGULATION PHENOTYPE

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Background. Disseminated intravascular coagulation (DIC) in patients with acute promyelocytic leukaemia (APL) typically manifests with severe haemorrhagic complications. While the coagulopathy of APL is notorious for its life-threatening bleeding risk, a purely thrombotic phenotype is rarely reported. We describe the case of a patient whose APL onset was characterized by widespread, multi-district arterial and venous thrombosis.

Case report. A 52-year-old woman, with a history of arterial hypertension and prior hysterectomy due to fibroids, presented to the Emergency Department with acute left-hand hyposthenia and hypoesthesia. Brain CT (non-contrast, angiography, and perfusion) and MRI confirmed a distal arterial occlusion of a branch of the right middle cerebral artery, associated with venous thrombosis of the right transverse sinus. Laboratory results showed pancytopenia, a mild increase in PT and reduced fibrinogen levels, confirming the diagnosis of DIC. A peripheral blood smear revealed 3% blasts, raising immediate suspicion of acute leukaemia. Treatment with ATRA was started promptly as soon as APL was suspected. Due to the extreme haemorrhagic risk of DIC associated with suspected APL, systemic thrombolysis was ruled out. After neurologic consultation, anticoagulation was initiated with unfractionated heparin (UFH) under rigorous aPTT monitoring. Subsequent total-body CT and Doppler ultrasound revealed a staggering thrombotic burden: thrombosis of the right hepatic vein and the portal branch for segment VIII and deep vein thrombosis (DVT) of the left popliteal vein. Sudden onset of left eye amaurosis, later confirmed by MRI as ischemia of the left retinal artery, resulting in permanent unilateral blindness. During hospitalization, a mild decline in platelet count prompted a switch from UFH to fondaparinux. Heparin-Induced Thrombocytopenia (HIT) was excluded via negative anti-heparin antibody testing. The diagnosis of APL was confirmed through both peripheral smear and bone marrow aspiration revealing the presence of promyelocytes and through FISH and PCR, showing detection of the t(15;17) translocation and the PML-RARα fusion transcript. As the patient was classified as non-high risk (WBC ≤ $10x10^9$/L), ATO was promptly added to the treatment regimen, with prednisone prophylaxis for differentiation syndrome. A good response to therapy was achieved in the absence of thrombotic or haemorrhagic complications, as well as serious adverse effects related to the treatment. At discharge, the patient was in morphological remission; therefore, anticoagulation was transitioned to apixaban, planned for a six-month full-dose course followed by a prophylactic dose (2.5 mg BID) until molecular remission is achieved.

Conclusions. DIC associated with hematologic malignancies is a pathologic condition characterized by increased fibrinolytic conditions and coagulation activation, rendering patients particularly prone to bleeding. This case represents a rare example of an exclusively thrombotic phenotype, which required a tailored therapeutic approach as the need for anticoagulation had to be carefully balanced against the high haemorrhagic risk. Given the lack of strong evidence regarding the optimal type of anticoagulant, the patient required close clinical and laboratory monitoring. Only after the acute phase was managed and the promyelocytic leukaemia treated, a DOAC was selected, a strategy supported by some case reports in the literature.