Massive pulmonary embolism presenting as disseminated intravascular coagulation

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Disseminated intravascular coagulation (DIC) can be defined as evidence of activation of the coagulation mechanism resulting in proteolysis of fibrinogen by thrombin and plasmin and an acute thrombocytopenia.

The association of pulmonary embolism (PE) with DIC has recently been reported but in reviewing recent textbooks of hematology, there is no mention of PE as a cause of DIC[1,7].

Clinicians need to be made aware of this association since it affects the patient who is thought to be autoanticoagulated as well as the patient who has DIC of unknown cause. PE needs to be included in the differential diagnosis of an autoanticoagulated state and in DIC of unknown etiology. In both instances the recommended treatment is full-dose intravenous heparin therapy[1,5,7].

Pulmonary embolism accounts for an estimated 300,000 hospitalizations and as many as 50,000 deaths annually[6]. Despite advances in diagnostic imaging it may go undetected until postmortem examination. Recognition of the association between DIC and pulmonary embolism may enhance the accuracy of the clinical diagnosis.

We report 2 cases of an unusual presentation of a massive PE and DIC.

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Case reports

Case 1

A 47-year-old Japanese man complained of a sudden onset of shortness of breath, hemoptysis and near loss of consciousness. Twelve days earlier he had undergone coronary artery bypass grafting.

He was known to have had diabetes mellitus and hypertension. Physical examination revealed left lung-field crackles. The admission chest roentgenogram demonstrated a small left pleural effusion. Laboratory tests were remarkable for thrombocytopenia. On arterial blood gas analysis, he had a widened A-a gradient.

A pulmonary angiogram demonstrated multiple bilateral pulmonary emboli. The patient subsequently developed bleeding into the thigh, thrombocytopenia, and other coagulation abnormalities. Fibrin split products (FSP) were increased, resulting in a decrease of the fibrinogen level. A bone-marrow aspiration was read as normal. A diagnosis of DIC was made. The patient recovered after treatment with full heparinization.

Case 2

An 80-year-old Caucasian woman with a history of steroid-dependent chronic obstructive pulmonary disease was admitted to hospital for treatment of *Pseudomonas pneumonia.* CBC and platelets were normal at admission.

Her medical history was also notable for congestive heart failure. Physical examination revealed dyspnea without cyanosis. The patient had a prolonged hospital course due to persistent weakness and shortness of breath. Primary hyperthyroidism was then diagnosed. An acute anemia developed. FSP were increased and the fibrinogen level was decreased. Acute thrombocytopenia developed and DIC was diagnosed. Heparin was not instituted given the presence of hematologic abnormalities. The patient died and at autopsy a massive, saddle, pulmonary embolus was found completely occluding both right and left lungs.

Discussion

Case 1 clinically illustrates DIC associated with a pulmonary embolus. Specifically, there was an acute thrombocytopenia and evidence of activation of the coagulation mechanism resulting in proteolysis of fibrinogen by thrombin and plasmin. This is the definition of DIC[7].

(Continued on page 125) ➤

The biochemical abnormalities can be detected by testing for circulating fibrin monomers and fibrin-degradation products. Once the occurrence of PE and DIC was recognized, treatment with heparin corrected the hematologic abnormalities and allowed the patient to recover.

Case 2 serves as a reminder that even in those clinical settings in which DIC can occur, PE may be present and should be considered even if laboratory examination suggests autoanticoagulation. The presence of an autoanticoagulation picture should not deter the clinician from treating with full doses of heparin, since the autoanticoagulation may represent DIC secondary to PE[1,5,6].

This association between PE and DIC is not well known[1,7]. The proposed mechanisms by which PE may result in DIC include: Liberation of tissue factors, endothelial damage, vascular malformation, decreased blood flow[7].

Stahl studied this association in 6 patients with DIC of unknown etiology and in each case he was able to document, by pulmonary angiogram or scanning, coexisting PE. Included were patients with DIC and DVT, DIC after aortic valve replacement, DIC after uncomplicated hemicolectomy for a benign villous adenoma, DIC and ulcerative colitis, DIC post-abdominal aortic surgery and DIC after a cesarean section. All cases were found to have coagulation abnormalites and evidence of DIC with thrombocytopenia. Full-dose heparin treatment resolved their hematologic abnormalities as well as their pulmonary emboli[1].

PE associated with an elevation of FSP was first described by Merskey. Elevated FSP was thought to represent the defibrination syndrome wherein fibrinolysis was the result of degradation and resolution of emboli[2]. Recent evidence suggests that PE itself activates the coagulation mechanism resulting in increased FSP and decreased fibrinogen levels; this activation is not expected to occur from fibrinolysis of an embolus.

Support for this concept comes from the finding that the fibrin formed after embolization has occurred is different from the fibrin in the original embolus itself. A study in dogs in which an I-125 fibrin-labeled autologous clot was embolized into the dog lung, revealed that the subsequent appearance of fibrin degradation products did not contain the radioactive label. Heparin given just prior to embolization was able to markedly inhibit the consumption of fibrinogen and the production of degradation products[8]. This suggests further evidence that DIC occurred secondary to PE; rather than fibrinolysis of the embolus.

This appears to be a specific activation that occurs in the lung; patients with thrombophlebitis or deep venous thrombosis give no evidence of DIC or of an increase in FSP unless there is also a PE[3,8]. In a study of 46 patients with suspected PE, the concentration of FSP when compared with the angiographic findings revealed that FSP was increased above 10 micrograms per milliliter in 19 patients with angiographically documented PE and were not elevated at all in the 22 patients

(Continued on page 134) ➤

MASSIVE PULMONARY *(Continued from page 125)*

that were negative for PE. There were 5 patients with equivocal findings[4].

Conclusion

Pulmonary embolism should be included in the differential diagnosis of DIC of unknown origin because prompt treatment with full-dose heparin will correct the hematologic abnormalities as well and may be lifesaving. An autoanticoagulated state does not always prevent the formation of thrombi, and it may obscure an occult PE. The presence of FSP may add to the understanding of the pathophysiology in PE and may be a laboratory clue that can assist in the management of both PE and DIC. This occurrence merits wider recognition by clinicians and further studies need to be conducted.

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