THE PROFIBROTIC ACTIVITY OF THE ANTI-FIBRYNOLITIC MARKERS IN THE BRONCHOALVEOLAR LAVAGE FLUID FROM PATIENTS WITH PROGRESSING IDIOPATHIC PULMONARY FIBROSIS

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The importance of coagulation and fibrinolysis systems have been implicated in the pathogenesis of idiopathic pulmonary fibrosis (IPF). However, available data are surprisingly scarce. Therefore, we investigated procoagulant (tissue factor, TF), fibrinolytic (urokinase type plasminogen activator, u-PA) and antifibrinolytic (plasminogen activator inhibitor-1, PAI-1 and PAI-2) activities in bronchoalveolar lavage fluids (BALF) from patients with active hypersensitivity pneumonitis (HP; n = 15), idiopathic pulmonary fibrosis (IPF, n = 15), and sarcoidosis (SARC, n = 20). IPF patients demonstrated highest levels of PAI-1 in BALF in comparison with the BBS and HP group ($P<0.002$), while uPA and TF activities were the lowest ($P<0.01$ and $P< 0.002$, respectively). The differences in PAI-2 levels did not reach significance. Importantly, the correlations between PAI-1 *vs.* DLco ($r=0.53$, $P<0.01$), TF *vs.* DLco ($r=-0.50$, $P<0.01$), TF *vs.* Cst ($r=-89$, $P<0.001$), and uPA *vs.* Cst ($r=0.46$, $P<0.01$) were observed in the IPF patients. In IPF, the BALF PAI-1 concentration correlated with the macrophage count ($r=0.61$, $P<0.01$), while the TF and uPA concentrations correlated with the overall cell count ($r=0.647$, $P<0.01$, $r=0.41$, $P<0.05$). Our study confirmed that increased procoagulant and antyfibrinolytic activities significantly contribute to the pathogenesis of IPF.