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Biomarkers identified for idiopathic pulmonary fibrosis

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The first evidence of a distinctive protein signature that could help to transform the diagnosis and improve the monitoring of the devastating lung disease idiopathic pulmonary fibrosis (IPF) is being reported by University of Pittsburgh School of Medicine researchers in this month's edition of *PLoS Medicine*, an open-access journal of the Public Library of Science.

In the paper, Naftali Kaminski, M.D., director of the Dorothy P. & Richard P. Simmons Center for Interstitial Lung Disease in the Division of Pulmonary, Allergy and Critical Medicine at the University of Pittsburgh School of Medicine, and his colleagues describe a unique combination of blood proteins that appears to distinguish IPF patients from normal controls with extraordinary sensitivity and precision.

"Our findings suggest that we may be able to monitor what is happening in the lungs by measuring certain proteins in the peripheral blood," explains senior author Dr. Kaminski, who also is associate professor of medicine. "More study is needed to confirm whether these biomarkers might be useful as a clinical blood test to detect lung fibrosis. But right now, there is no straightforward test for IPF. The lung is not highly accessible; biopsy procedures carry risk, and while imaging is good, it can't follow the disease biologically."

IPF is a degenerative illness distinguished by progressive lung scarring and diminished breathing capacity, typically leading to death within about five years of diagnosis. It is estimated that 5 million people

worldwide and 130,000 in the United States are affected by pulmonary fibrosis and about 30,000 people die of the disease every year.

For this study, researchers analyzed the concentrations of 49 proteins in the plasma of 74 patients with IPF and 53 normal controls. A combination of five proteins related to normal tissue breakdown and remodeling and certain disease processes, including arthritis and cancer, was found to be highly indicative of IPF.

Increases in two of the five, matrix metalloproteinases (MMP) 7 and 1, also were observed in tissue and fluid taken from the lungs of IPF patients. Other proteins in the IPF signature are matrix metalloproteinase 8, insulin-like growth factor binding protein 1 and tumor necrosis factor receptor superfamily member 1A.

“These proteins were increased in IPF patients, but not in patients with lung illnesses such as chronic obstructive pulmonary disease,” says Ivan O. Rosas, M.D., first author on the study and assistant professor of medicine, University of Pittsburgh School of Medicine. Elevated MMP1 and MMP7 also distinguished IPF when compared to levels associated with another disease that closely mimics IPF, called subacute/chronic hypersensitivity pneumonia. In particular, increased concentrations of MMP7 “may be indicative of asymptomatic lung disease and perhaps reflect disease progression,” Dr. Rosas says.

“One of the challenges is to know whether a blood protein actually reflects the situation in the lung,” notes Thomas J. Richards, Ph.D., study co-first author and research assistant professor in the Division of Pulmonary, Allergy and Critical Care Medicine, University of Pittsburgh School of Medicine. The team evaluated all the genes expressed in IPF-affected lung tissue to determine the proteins in the peripheral blood on which they should focus. Based on their detailed analysis, the team believes that increased levels of these five proteins probably are

reflective of the disease.

“IPF can have a slow progression, so drug companies may wait a long time to see whether a particular drug is having any effect,” says Dr. Kaminski. “But a blood biomarker could indicate whether a drug is working earlier. The biomarkers also might be used for risk assessment and for evaluation of disease progression.”

Some known causes of pulmonary fibrosis include occupational and environmental exposure to asbestos, metal dust, farming chemicals and mold, an inflammatory disease called sarcoidosis, radiation, drug reactions, autoimmune disorders and possibly a genetic predisposition, according to the American Lung Association.

Most cases are considered to be idiopathic, or of unknown origin. There is no proven effective therapy for IPF, and most drug interventions are considered experimental. Long-term benefit may be possible with lung transplantation, a radical approach dependent upon a limited number of donated organs.

Source: University of Pittsburgh

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