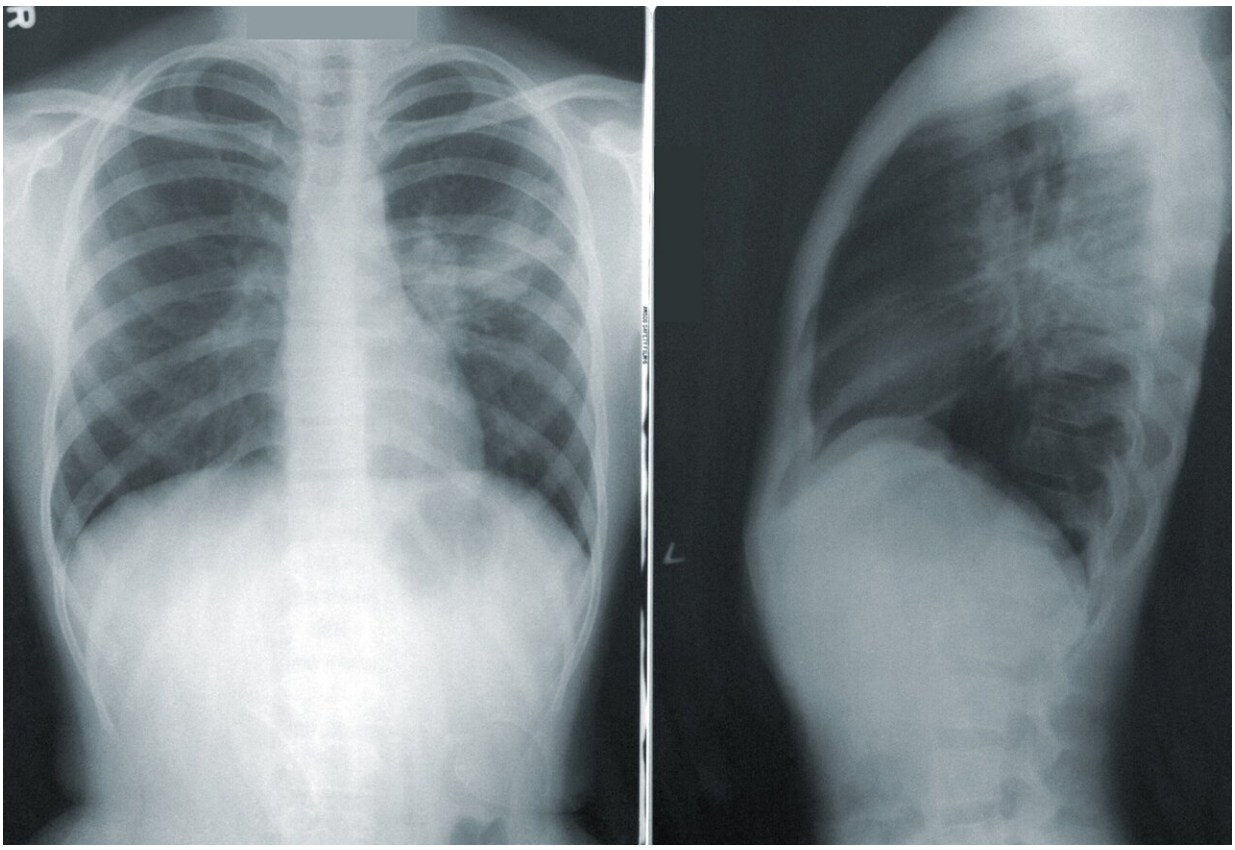
Study offers insight into management of patients who have interstitial pneumonia with autoimmune features

July 5 2022



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Interstitial pneumonia with autoimmune features (IPAF) is a disease

with many possible causes and no standard of care, making it particularly difficult to treat. While immunosuppressant drugs are primarily prescribed, they don't work for all patients.

A new study of patients treated for IPAF at UT Southwestern offers insight into the disease, which can result in a reduced quality of life and sometimes early death. Among their findings, the researchers discovered that a combination therapy of mycophenolate mofetil and prednisone was associated with a lack of disease progression. The study was published in the *Journal of Clinical Rheumatology*.

"Some patients with IPAF don't require any treatment at all and some seem to not respond to immunosuppression. Clinicians are now finding that a new class of medications called antifibrotics may be an appropriate treatment, but we still don't know which IPAF patients need to be treated with immunosuppression and which need to be treated with antifibrotics or both," said Elena Joerns, M.D., a postdoctoral fellow in the Division of Rheumatic Diseases at UT Southwestern, who led the study with Una Makris, M.D., Associate Professor of Internal Medicine and Population and Data Sciences.

Drs. Joerns and Makris, working with colleagues from the Division of Rheumatic Diseases and the Division of Pulmonary and Critical Care Medicine, sought to define features of IPAF that would predict which patients will respond favorably to immunosuppression. Crucially, the identification of such features would help to guide physicians on the best treatment for their patients.

To this end, the researchers reviewed medical records of 63 patients with IPAF. They evaluated clinical, serologic, and morphologic disease features, as well as demographic characteristics. The group did not find any characteristics associated with a response to immunosuppression.

"Although we did not find statistically significant differences, we did identify trends that could be informative for future studies. For example, no patient positive for anti-synthetase antibody progressed and there was greater progression among men and former or current smokers, which aligns with findings from other studies," said Dr. Makris. Moreover, the study found that immunosuppressive therapy with mycophenolate mofetil and prednisone was associated with lack of disease progression in immunosuppressed IPAF patients, including those with usual interstitial pneumonia pattern, a feature suggested by some cohorts to be predictive of worse mortality.

The study was limited by a small cohort of patients from a single center. Additionally, the characteristics that could be evaluated were limited to those in the medical records. Dr. Joerns explained that patients do not all receive the same battery of tests when diagnosed. More in-depth serologic workups of IPAF patients may be needed to improve diagnoses and identify features predictive of response to immunosuppression.

The study results underscore the challenges in treating IPAF and the mystery that surrounds this newly defined disease.

"There is still a lot to learn about IPAF so that we can optimally manage this patient population," said Dr. Joerns, who is pursuing further research. "But we have an excellent collaboration at UT Southwestern between Rheumatology, Pulmonology, Radiology, and Pathology, all of whom are essential for classifying and managing these patients, which makes our center well poised to answer these questions on the pathogenesis of IPAF."

More information: Elena K. Joerns et al, Variables Associated With Response to Therapy in Patients With Interstitial Pneumonia With Autoimmune Features, *JCR: Journal of Clinical Rheumatology* (2021). DOI: 10.1097/RHU.0000000000001808

Provided by UT Southwestern Medical Center

Citation: Study offers insight into management of patients who have interstitial pneumonia with autoimmune features (2022, July 5) retrieved 19 April 2024 from https://medicalxpress.com/news/2022-07-insight-patients-interstitial-pneumonia-autoimmune.html

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