

Cystic Fibrosis Research News

Title:

Fibrocyte accumulation in the lungs of cystic fibrosis patients

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What was your research question?

Fibrocytes are a unique cell type that originates from bone marrow. These cells display features of both immune cells and collagen producing cells called fibroblasts. We assessed whether fibrocytes accumulate in the distal or gas exchange areas of the lung and identified alterations in the gene expression profiles in patients with CF lung disease.

Why is this important?

Fibrosis has been identified in the lungs of patients with end-stage CF lung disease. Fibrosis is responsible for decline in lung function and ultimately causes death. During fibrosis, fibroblasts produce excess collagen and form scar tissue in the lungs. Fibrocytes accumulate to form lesions that have been linked to declining lung function. Therefore, identifying fibrocytes in the lung and characterizing their association with CF lung disease may provide insights on disease progression and identify potential treatment options.

What did you do?

To study fibrocyte accumulation, we quantified the number of fibrocytes in distal lung tissue in healthy lungs compared to CF lungs with end-stage disease. We quantified fibrocytes in the airway fluid of CF patients. We also used cell culture methods to isolate fibrocytes from the lungs of CF patients and healthy subjects and identify dysregulated genes. We utilized



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bioinformatics tools as well to identify altered biological processes in fibrocytes that accumulate in CF-patient lungs.

What did you find?

We identified increased fibrocytes in the distal areas of CF lungs with end-stage lung disease compared to tissue from healthy control lungs. We also detected fibrocytes in the airway fluid of CF patients. Gene expression profiles that regulate the expression of collagen were altered in fibrocytes from CF lungs compared to healthy controls.

What does this mean and reasons for caution?

Our findings show the accumulation of fibrocytes around the functional units of CF lungs with the end-stage disease compared to healthy controls. As we only used CF lungs with end-stage disease, we were unable to assess their role during the progression of disease. Also, the lack of a healthy control group to quantify fibrocytes in airway fluid makes it difficult to determine if fibrocytes are normally detectable in airway specimens from healthy lungs. We must interpret the gene expression data pertaining to CF fibrocytes cautiously as the study used cultured fibrocytes and culture conditions may impact the biology of fibrocytes.

What's next?

These findings will support studies examining whether fibrocyte accumulation in airway fluid or blood is associated with collagen deposition and lung function decline in CF patients. Identifying the role of fibrocytes in collagen deposition could identify new avenues for the development of anti-fibrotic therapies for CF patients.

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