First study to examine role of mast cells in cystic fibrosis

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Dr. Tong-Jun Lin is conducting groundbreaking research into cystic fibrosis, a fatal inherited disease. His study is the first in the world to examine the role of a special type of cell, called mast cells, in airway inflammation in CF patients. If it is determined that mast cells are an important factor, then medications may be developed to help control the inflammation. An effective treatment could vastly improve the health and quality of life of patients with this disease. His preliminary research is promising, but additional studies need to be conducted.

In cystic fibrosis, an unusually thick, sticky mucus clogs the bronchial tubes (airway passages) in the lungs and blocks the ducts, or exit passages, from the pancreas and intestines, so these organs cannot function properly In the lungs, these secretions clog the airways and lead to chronic obstruction, inflammation and infection. The secretions also prevent the proper digestion of food, forcing people with CF to take artificial enzymes – as many as 40 pills a day – to aid in digestion. As well, daily physiotherapy is required to help free the lungs of congestion.

Progressive lung disease is the predominant cause of illness and death in CF patients. One in every 25 Canadians carries the gene for CF, and it is estimated that one in every 2,500 children born in Canada has the disease. Since Canadian researchers discovered the gene responsible for CF in 1989, research has greatly lengthened the lifespan of people with CF. In 1960, a child with cystic fibrosis rarely lived to the age of four, now people with CF live productively into their mid-30s and beyond.

CF is characterized by airway obstruction, chronic respiratory tract infections and excessive inflammation. Pseudomonas aeruginosa (P aeruginosa) is the most common bacterium infecting the CF lung, colonizing (inhabiting long term) the lungs of people with CF before they reach the age of 10. It is ubiquitous in several diseases and noted for its resistance to antibiotics. Also noted for its environmental versatility and its ability to cause disease in those who are susceptible, P aeruginosa can cause extensive tissue damage and interfere with immune system defenses. The body's response to P aeruginosa includes inflammation, which causes repeated episodes of intense breathing problems. Mast cells play an essential role in inducing inflammatory and immediate allergic reactions. They release potent inflammatory mediators (such as histamine, proteases, and cytokines) that act on blood vessels, smooth muscle connective tissue, mucous glands, and inflammatory cells.

The role of mast cells in P aeruginosa-induced inflammation has not been studied previously. Dr. Lin's work provides new information on the mechanisms of P aeruginosa infection. He and his research team found that mast cells produce significant amounts of various cytokines and chemokines after P aeruginosa stimulation.

The results of Dr. Lin's work suggest that mast cells may potentially serve as a target for the development of drugs and therapeutic approaches for the treatment P aeruginosa-involved diseases, such as cystic fibrosis. Additional studies now need to be conducted to better understand the role of mast cells in P aeruginosa-induced inflammation.

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Research Results