MedicalXpress

Researchers elucidate a molecular mechanism associated with an immune disorder

June 7 2012

A team of researchers at the IRCM led by Dr. André Veillette made an important breakthrough in the field of immunology, which will be published online today by the scientific journal *Immunity*. The scientists explained a poorly understood molecular mechanism associated with a human immune disorder known as XLP disease or Duncan's syndrome.

"We studied the SAP molecule, which plays a critical role in multiple different types of immune cells," says Dr. Veillette, Director of the Molecular Oncology research unit at the IRCM. "More specifically, we wanted to understand why SAP is an essential component of natural killer cells' ability to eliminate abnormal blood cells."

Natural killer (NK) cells are crucial to the immune system, and provide rapid responses against cancer and virus-infected cells, especially blood cells as can be found in leukemia and lymphomas. Patients with XLP are at a high risk of developing lymphomas.

"Until now, the way by which SAP enhances NK cells' response to abnormal blood cells was not well understood," explains Zhongjun Dong, former researcher in Dr. Veillette's laboratory and first author of the article. "We discovered that SAP stimulates the function of NK cells through a double mechanism. On one hand, it couples the necessary genes and enzymes to increase NK cell responses, and on the other hand, it prevents genes from inhibiting these responses." Dr. Dong is now a

professor at Tsinghua University, a leading university in China.

"The SAP molecule is important in immunity, as it is associated with most cases of XLP disease," adds Dr. Veillette. "In addition, our findings may have implications on the role of SAP in other diseases such as lupus and arthritis."

According to the XLP Research Trust, X-linked lymphoproliferative disease (XLP), also known as Duncan's syndrome, is a fatal disease affecting boys worldwide. The cause of the condition was only found in 1998, so many cases may not yet have been properly diagnosed. If untreated, approximately 70% of patients with XLP die by the age of 10.

Dr. Veillette's research is funded by the Canada Research Chairs program and the Canadian Institutes for Health Research (CIHR). "I applaud Dr. Veillette and his team for their research in the field of human immune disorder and their breakthrough discovery in understanding the role of the SAP protein in controlling abnormal blood cells," said Dr. Marc Ouellette, Scientific Director of the CIHR Institute of Infection and Immunity. "Their work will contribute to a better understanding of our immune system and how to treat human immune diseases for improved health for all Canadians."

More information: For more information on this discovery, please refer to the article summary published online by Immunity: www.cell.com/immunity/abstract … 1074-7613(12)00231-2

Provided by Institut de recherches cliniques de Montreal

molecular-mechanism-immune-disorde.html