The challenge of diagnosing Langerhans cell histiocytosis

One of the challenges involved in diagnosing Langerhans cell histiocytosis (LCH) is that the disease can present in so many different ways, according to Dr. Bruce Crooks, Assistant Professor of Paediatric Haematology/Oncology & Medical Education at the IWK Health Centre. Dr. Crooks is also the principal investigator of a Canadian Paediatric Surveillance Program (CPSP) study on LCH, which will help raise awareness and global knowledge of LCH.

LCH, involves a proliferation of pathogenic Langerhans cells. It can present as a rash that looks like napkin dermatitis or as a bony lesion, affecting the skull or long bones. In severe forms, the disease can involve many tissues, including lungs, liver, bone marrow and the central nervous system.

Another challenge of diagnosing this disease is that it's so rare, with likely only 30 to 40 cases a year in Canada. "Doctors are not primed; they are not thinking about it," explains Dr. Crooks. "LCH puzzles many doctors. You need to have heard of it to even think about it."

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>
> Dr. Bruce Crooks

Early diagnosis is key

If LCH is caught early, treatment can be relatively straightforward.

"Sometimes just an injection of steroids can work," says Dr. Crooks. However, even though it is not a cancer (the cells are not malignant), LCH can behave like one. As a result, treatment can also involve surgery and/or chemotherapy. As the disease progresses, it can cause long-term health effects, for example by eroding into the bone, head or eye.

Before LCH is diagnosed, patients may be referred to services such as orthopaedics, neurosurgery, otorhinolaryngology, dermatology and endocrinology for treatment.

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Getting a clearer picture of LCH

The study also aims to identify the epidemiological features of LCH in Canada so that diagnosis and management can be improved, areas for further research clarified, and resources for LCH patients more effectively allocated.

It is also unclear whether LCH varies among ethnic groups. The CPSP study may help to shed some light on this question simply because of the diverse nature of Canada's population.

The study began in July 2009 and will run until June 2011. Clinically active paediatricians will be surveyed, along with haematologists/oncologists and other allied specialty physicians.

"I am hopeful that the study results will help us make sure we don't miss cases—and that all cases are treated the way they should be," says Dr. Crooks.