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Immunodeficiency Diseases- Part 2

In the last article, we have learnt how the immune system works. In today's article, we will learn how things could go wrong and what the impact might be.

When do you suspect immunodeficiency?

All of us get infections ranging from common cold to pneumonia from time to time. It does not mean we have immunodeficiency. Young children with naïve immune system get upper respiratory infections and ear infections often. As they get older, their immune system matures to a degree that they stop getting these infections. When do we suspect problems with our immune system then? According to the Immune Deficiency Organization, a nonprofit organization devoted to helping patients and families with immunodeficiency, an immune deficient disease should be suspected if you get infections that are recurrent (keep coming back), persistent (won't completely clear up or clears very slowly), severe (requires hospitalization or intravenous antibiotics), unusual (caused by an uncommon organism) and or runs in the family (other family members have had a similar susceptibility to infections).

Some of the warning signs of immunodeficiency include two or more serious sinus infections within 1 year, two or more months on antibiotics with little effect, two or more pneumonias within 1 year, failure of an infant to gain weight or grow normally, recurrent, deep skin or organ abscesses, persistent thrush in mouth or elsewhere on skin, after age 1, need for intravenous antibiotics to clear infections, two or more deep-seated infections or a family history of Primary Immunodeficiency Disease.

Immunodeficiency diseases also predispose individuals to autoimmune diseases such as lupus, rheumatoid arthritis, Sjögren's syndrome etc. and to cancers and lymphoma. Therefore if you have strong family history of autoimmune disorders or cancers/ lymphoma and get frequent and severe infections, then you should get checked for immunodeficiency. Children with small or absent tonsils and lymph nodes may have primary immunodeficiency as well.

Types of Immunodeficiency

Immune deficiency diseases are broadly classified into primary and secondary forms. Secondary forms of immunodeficiency are much more common and include such well known disorders as HIV/AIDS. Besides HIV, conditions such as cancer, chemotherapy,

radiation treatment, malnutrition, some medications and chronic diseases affecting liver, kidneys and intestines could lead to secondary forms of immunodeficiency. In these conditions, the immune system is weakened by the underlying disease and adequate treatment of the original problem will improve the immune function.

In contrast there are well over 100 forms of primary immune deficiency (PID) disorders described by the World Health Organization. They are usually rare, can affect all ages and both sexes, and usually affect several members in the same family. Some of the diseases are so rare that an average physician may not see one in a life-time. On the other hand, IgA deficiency- a form of PID affecting IgA antibody protein affects 1 in 700 people. The primary immunodeficiency diseases are genetically acquired. Some of them have well-defined clinical presentation and course and are well amenable to diagnosis and treatment. Others are difficult to diagnose and treat. For some, no specific treatment is available.

How is immune deficiency diagnosed?

A high level of suspicion is required to diagnose immune deficiency disorders. History as above gives the most important clue. Physical examination by an immunologist may strengthen the suspicion. Failure to thrive (failure to gain weight and height), scarred ear drums and draining ears, signs of chronic sinusitis, small or absent tonsils and lymph glands, presence of oral thrush, sore and bleeding gums, malformed blood vessels in the conjunctiva and skin, abnormal breath sounds on listening to lungs, finger clubbing (convex finger nails), severe eczema, bleeding spots in the skin, heart murmur and enlarged liver and spleen are some of the findings to look for on physical examination.

If the history and physical examination suggest PID as a possibility, then your doctor may order blood tests to clinch the diagnosis. This includes doing a full blood count, measuring the levels of different antibody proteins in the blood, checking to see if your body made adequate antibody response to some of previously given vaccines (Tetanus and Pneumonia shots) or naturally acquired diseases and measuring the level of complement proteins. If the blood tests reveal the presence of immune deficiency, your immunologist will order more sophisticated tests to narrow down the diagnosis.

In the next article, we will learn about how the immune deficiency disorders are treated at present and latest developments in their treatment.

About the author:

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