12.6B: Secondary Immunodeficiency Diseases

Secondary immunodeficiencies refer to acquired immune system disorders.

Learning Objectives

- Define immunodeficiency and list the types that occur

Key Points

- A deficit in the immune system can lead to unusually severe or uncommon recurrent infections.
- Secondary immune deficiencies or acquired deficiencies, more frequent than primary immune deficiencies, are problems of the immune system that are not genetic and which are caused by external factors.
- Secondary immunodeficiency disorders can occur in, for example, malnutrition, aging, many types of cancer (such as leukemia, lymphoma, multiple myeloma), and certain chronic infections such as acquired immunodeficiency syndrome (AIDS).
- Immunosuppression is one form of secondary immunodeficiency performed to prevent the body from rejecting an organ transplant, treating graft-versus-host disease after a bone marrow transplant, or for the treatment of autoimmune diseases, such as rheumatoid arthritis or Crohn's disease.
- A person who is undergoing immunosuppression or whose immune system is weak for other reasons (for example, chemotherapy, HIV, and Lupus), is said to be immunocompromised.

Key Terms

- **immunodeficiency**: A depletion in the body’s natural immune system, or in some component of it.
• **immunocompromised**: Having an immune system that has been impaired by disease or treatment.
• **immunosuppressive**: Having the capability to suppress the immune system, capable of immunosuppression.
• **secondary infection**: any infection that arises subsequent to a pre-existing infection; but especially a nosocomial infection.

Immunodeficiency (or immune deficiency) is a state in which the immune system’s ability to fight infectious disease is compromised or entirely absent. Immunodeficiency may also decrease cancer immunosurveillance. Most cases of immunodeficiency are acquired (“secondary”) but some people are born with defects in their immune system, or primary immunodeficiency. Transplant patients take medications to suppress their immune system as an anti-rejection measure, as do some patients suffering from an over-active immune system. A person who has an immunodeficiency of any kind is said to be immunocompromised. An immunocompromised person may be particularly vulnerable to opportunistic infections, in addition to normal infections that could affect everyone. Distinction between primary versus secondary immunodeficiencies are based on, respectively, whether the cause originates in the immune system itself or is, in turn, due to insufficiency of a supporting component of it or an external decreasing factor of it.

**Figure: Main Symptoms of AIDS**

Aquired immunodeficiency syndrome (AIDS) is defined in terms of either a CD4+ T cell count below 200 cells per µL or the occurrence of specific diseases in association with an HIV infection. In the absence of specific treatment, around half the people infected with HIV develop AIDS within 10 years. The most common initial conditions that alert to the presence of AIDS are pneumocystis pneumonia and cachexia.

**Primary Immunodeficiency (PID)**

A number of rare diseases feature a heightened susceptibility to infections from childhood onward. Primary Immunodeficiency is also known as “congenital immunodeficiencies.” Many of these disorders are hereditary and are
autosomal recessive or X-linked. There are over 80 recognized primary immunodeficiency syndromes; they are generally grouped by the part of the immune system that is malfunctioning, such as lymphocytes or granulocytes. The treatment of primary immunodeficiencies depends on the nature of the defect and may involve antibody infusions, long-term antibiotics, and (in some cases) stem cell transplantation.

### Secondary Immunodeficiencies

Secondary immunodeficiencies, also known as acquired immunodeficiencies, can result from various immunosuppressive agents, for example, malnutrition, aging and particular medications (e.g., chemotherapy, disease-modifying antirheumatic drugs, immunosuppressive drugs after organ transplants, glucocorticoids). For medications, the term immunosuppression generally refers to both beneficial and potential adverse effects of decreasing the function of the immune system, while the term immunodeficiency generally refers solely to the adverse effect of increased risk for infection. Many specific diseases directly or indirectly cause immunosuppression. This includes many types of cancer, particularly those of the bone marrow and blood cells (leukemia, lymphoma, multiple myeloma), and certain chronic infections. Immunodeficiency is also the hallmark of acquired immunodeficiency syndrome (AIDS), caused by the human immunodeficiency virus (HIV). HIV directly infects a small number of T helper cells and also impairs other immune system responses indirectly.