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1. Identify and briefly explain 5 primary immunodeficiency disorders
2. **Autoimmune Lymph proliferative Syndrome(ALPS):** Is an inherited disorder in which the body cannot properly regulate the number of immune system cells(lymphocytes). ALPS is characterized by the production of an abnormally large number of lymphocytes (lymph proliferation) .Accumulation of excess lymphocytes results in enlargement of the lymph nodes (lymphadenopathy), the liver (hepatomegaly) and the spleen (splenomegaly). Autoimmune disorders also occur in ALPS. Autoimmune disorders occur when the immune system malfunction and attack the body’s tissues and organs. Most of the autoimmune disorders associated with ALPS target and damage blood cells. For example the immune system may attack red blood cells (autoimmune hemolytic anemia) white blood cells (autoimmune neutropenia) or platelets (autoimmune thrombocytopenia). Less commonly autoimmune disorders that affect other organs and tissues occur in people with ALPS. These disorders can damage the kidneys (glomerulonephritis), Liver (autoimmune hepatitis), eyes(uveitis), or nerves(Guillain-Barre syndrome). Skin problems, usually rashes or hives (urticarial), can also occur in ALPS. People with classic form of ALPS generally have a normal-life span, but have a greatly increased risk of developing cancer of the immune system cells (lymphoma) compared with general population.
3. **Chronic granulomatous disease:** Is an inherited primary immunodeficiency disease which increases the body’s susceptibility to infection caused by certain bacterial and fungi. Granulomas are masses of immune cells that form at sites of infection or inflammation. Its occurs when a type of white blood cells (phagocyte) that usually help to fight infections do not work properly. As a result the phagocytes cannot protect your body from bacterial and fungi infection. People with chronic granulomatous disease may develop infection in the lungs, skin, lymph nodes, liver, stomach and intestines, or other areas. They may develop clusters of white blood cells in infected areas. Most people are diagnosed with CGD during childhood, but some people may not be diagnosed until adulthood. CGD makes it difficult for cells to called neutrophils to produce hydrogen peroxide to fight specific kinds of bacterial and fungi. People with CGD are generally healthy until they become infected with one of these germs. The severity of this infection can lead to prolonged hospitalization for treatment. Children with CGD are often healthy at birth but develop severe infection in infancy or early childhood. The most common form of CGD is genetically inherited in an x-linked manner, meaning it only affect boys. There are also autosomal recessive forms of CGD that affects both sexes.
4. **Common variable immunodeficiency**: Is an antibody deficiency that leaves the immune system unable to defend against bacteria and viruses, resulting in recurrent and often severe infection primarily affecting the ears, sinuses, and respiratory tract (sinopulmonary infection. CVID can be associated with autoimmune disorders that affect other blood cells causing low numbers of white blood cells or platelets, anemia, arthritis and other condition such as endocrine disorders. Gastrointestinal problems including chronic diarrhea, weight loss, nausea, vomiting and abdominal pain can also be present. In some forms of CVID patients develop granulomas in the lungs, lymph nodes, liver, skin other organs. People with are also at an increased risk for certain cancers (Lymphoid and gastrointestinal cancers primarily). CVID has an autoimmune disorder which occurs in the immune system malfunctions and attack the body’s tissues and organs. The blood cells are most frequently affected by autoimmune attack in CVID; the most commonly occurring autoimmune disorders are immune thrombocytopenia which is an abnormal bleeding disorder caused by decreased in cells involved in blood clotting called platelets, and autoimmune hemolytic anemia, which results in premature destruction of red blood cells.
5. **Congential neutropenia syndrome**: Is a condition that causes affected individuals to be prone to recurrent infections. People with this condition have shortage (deficiency) of neutrophils, a type of white blood cells that plays a role in inflammation and in fighting infection. The deficiency of neutrophils, called neutropenia is apparent at birth or soon afterwards. It leads to recurrent infection beginning in infancy, including infection of the sinuses, lungs, and the liver. Affected individuals can also develop fevers and inflammation of the gums (gingivitis) and skin. Approximately 40percent of affected persons have decreased bone density. Approximately 20 percent of people with severe congenital neutropenia also develop certain cancerous condition of the blood, particularly myelodysplastic syndrome or leukemia during adolescence. Some people with severe congential neutropenia have additional health problems such as seizures, developmental delay, or heart and gential abnormalities.
6. **DOCK8 deficiency**: Is a rare immune disorder named after the mutated gene responsible for the disease. It is the autosomal recessive form of hyperimmunogloblin E syndrome, a genetic disorder characterized by elevated immunoglobulin E levels, eosinophilla, and recurrent infection with staphylococcus and viruses. It is caused by a mutation in the DOCK8 gene. People with this syndrome have lower than normal numbers of immune cells, which have a diminished capacity to move through dense tissues like the skin. These abnormalities lead to recurrent viral infection of the skin and respiratory system. People with DOCK8 deficiency also typically have allergies, asthma, and increased risk for some types of cancer. DOCK8 deficiency is associated with high levels of an antibody called immunoglobulin E, or IgE. The syndrome formerly was known as hyper-IgE syndrome, or ARHIES. DOCK8 deficiency is one of many hyper-IgE syndromes.
7. Identify and briefly explain 2 secondary immunodeficiency disorders
8. **Multiple myeloma(cancer of the plasma cells, which produce antibodies):** Multiple myeloma is a cancer that forms in a type of white blood cells called plasma cells. Plasma cells help to fight infection by making antibodies that recognize and attack germs. Multiple myeloma causes cancer cells to accumulate in the bone marrow, where they crowd out healthy blood cells. Rather than produce helpful antibodies, the cancer cells produce abnormal proteins that can cause complications. Is a plasma cell malignancy in which monoclonal plasma cells proliferate in bone marrow, resulting in an overabundance of monoclonal Para protein (M protein), and destruction of bone and displacement of other hematopoietic cell lines.
9. **AIDS (Acquired immune deficiency syndrome):** Is a disease that can develop in people with HIV. It is the most advanced stage of the HIV. Untreated, HIV can progress to AIDS within decade. There is no cure for AIDS and without treatment. Life expectancy after diagnosis is about three years. This may be shorter if the person develops severe opportunistic illness. If a person’s CD4 count falls below 200, then the person have AIDS. When the immune system is very weak people with AIDS get serious infection and health problems. It is also as results of advanced HIV infection which has destroyed the immune system.