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Assignment Title: Immunodeficiency Disorders

Primary Immunodeficiency Disorder

1.Chronic Granulomatous Disease: Chronic granulomatous disease (CGD) is a genetic disorder in which white blood cells called phagocytes are unable to kill certain types of bacteria and fungi. People with CGD are highly susceptible to frequent and sometimes life-threatening bacterial and fungal infections. CGD is caused by defects in an enzyme, NADPH oxidase, that phagocytes need to kill certain bacteria and fungi. Mutations in one of five different genes can cause these defects.

People with CGD are highly susceptible to infections caused by certain bacteria and fungi, such as Staphylococcus aureus, Serratia marcescens, Burkholderia cepacia, Nocardia species, and Aspergillus species. These people may develop abscesses (boils) in their lungs, liver, spleen, bones, or skin; and masses of cells, called granulomas, that can obstruct the bowel or urinary tract.

2.Congenital Neutropenia Syndromes: Congenital neutropenia syndromes are a group of disorders present from birth that are characterized by low levels of neutrophils, a type of white blood cell necessary for fighting infections. People with congenital neutropenia experience bacterial infections early in life. These may cause inflammation of the umbilical cord stump, abscesses (or boils) on the skin, oral infections and pneumonia.Congenital neutropenia also increases one’s risk for developing myelodysplastic syndromes (MDS), blood disorders that are distinguished by low levels of various blood cells. MDS may progress to a type of blood-cell cancer called acute myeloid leukemia.

3.Autoimmune Lymphoprofilerative Syndrome: Autoimmune lymphoproliferative syndrome (ALPS) is a rare immune disorder first described by NIH scientists in the mid-1990s that can cause numerous autoimmune problems, such as low levels of red blood cells, clot-forming platelets, and infection-fighting white blood cells. These problems can increase the risk of infection and hemorrhage. Most cases of ALPS are caused by mutations in the FAS gene. FAS produces a receptor that, when activated, leads to programmed cell death, or apoptosis. This programmed death is an important part of the normal cell lifecycle. When cells do not receive the message that it is time for them to die, an abnormal buildup of cells can result. In the case of ALPS, mutations in FAS cause an abnormal buildup of white blood cells. There currently is no standard cure for ALPS. The disorder can be managed by treating low blood-cell counts and autoimmune diseases that occur in people with ALPS, as well as by monitoring for and treating the proliferation of immune cells, enlarged spleen, and lymphoma.

4.Leukocyte Adhesion Deficiency: Leukocyte adhesion deficiency (LAD) is a rare, inherited immune disorder in which immune cells called phagocytes are unable to move to the site of an infection to fight off invading pathogens. People with LAD experience recurrent, life-threatening infections and poor wound healing. LAD is caused by a mutation in the gene ITGB2, which provides instructions for the phagocyte surface molecule CD18. Treatments for LAD include antibiotics to prevent and treat infection and, in some cases, bone marrow transplants from a healthy donor.

5. Interferon Gamma, Interleukin 12 and Interleukin 23 Deficiencies:Interferon gamma, interleukin 12 and interleukin 23 deficiencies are rare, inherited immune disorders in which the body fails to produce one or more of these signaling molecules, which allow infection-fighting immune cells to communicate. Deficiencies in these molecules lead to increased susceptibility to bacterial and viral infections. Many people with these deficiencies develop granulomas, or inflammatory lesions that form in tissues and organs because of recurring infections. While many of these deficiencies begin to cause symptoms in infancy or childhood, some symptoms appear later in life. Treatment includes antibiotic therapy to prevent infections and, in some cases, bone marrow transplant from a healthy donor.

Secondary Immunodeficiency Disorder

1.AIDS

AIDS is a disease that can develop in people with HIV. It’s the most advanced stage of HIV. But just because a person has HIV doesn’t mean they’ll develop AIDS.

HIV kills CD4 cells. Healthy adults generally have a CD4 count of 500 to 1,500 per cubic millimeter. A person with HIV whose CD4 count falls below 200 per cubic millimeter will be diagnosed with AIDS.

A person can also be diagnosed with AIDS if they have HIV and develop an opportunistic infection or cancer that’s rare in people who don’t have HIV. An opportunistic infection, such as pneumonia, is one that takes advantage of a unique situation, such as HIV.

Untreated, HIV can progress to AIDS within a decade. There’s no cure for AIDS, and without treatment, life expectancy after diagnosis is about three yearsTrusted Source. This may be shorter if the person develops a severe opportunistic illness. However, treatment with antiretroviral drugs can prevent AIDS from developing.

If AIDS does develop, it means that the immune system is severely compromised. It’s weakened to the point where it can no longer fight off most diseases and infections. That makes the person vulnerable to a wide range of illnesses, including:

•pneumonia

•tuberculosis

•oral thrush, a fungal infection in the mouth or throat

•cytomegalovirus (CMV), a type of herpes virus

•cryptococcal meningitis, a fungal infection in the brain

•toxoplasmosis, a brain infection caused by a parasite

•cryptosporidiosis, an infection caused by an intestinal parasite

•cancer, including Kaposi’s sarcoma (KS) and lymphoma.

 The shortened life expectancy linked with untreated AIDS isn’t a direct result of the syndrome itself. Rather, it’s a result of the diseases and complications that arise from having an immune system weakened by AIDS.

2.Leukemia

Leukemia is a cancer of the blood or bone marrow. Bone marrow produces blood cells. Leukemia can develop due to a problem with blood cell production. It usually affects the leukocytes, or white blood cells.Leukemia is most likely to affect people over the age of 55 years, but it is also the most common cancer in those aged under 15 years. Acute leukemia develops quickly and worsens rapidly, but chronic leukemia gets worse over time. There are several different types of leukemia, and the best course of treatment and a person’s chance of survival depends on which type they have.

 Leukemia develops when the DNA of developing blood cells, mainly white cells, incurs damage. This causes the blood cells to grow and divide uncontrollably.Healthy blood cells die, and new cells replace them. These develop in the bone marrow.The abnormal blood cells do not die at a natural point in their life cycle. Instead, they build up and occupy more space.

 As the bone marrow produces more cancer cells, they begin to overcrowd the blood, preventing the healthy white blood cells from growing and functioning normally.

 Risk factors

There is a range of risk factors for leukemia. Some of these risk factors have more significant links to leukemia than others:

Artificial ionizing radiation: This could include having received radiation therapy for a previous cancer, although this is a more significant risk factor for some types than others.

Certain viruses: The human T-lymphotropic virus (HTLV-1) has links to leukemia.

Chemotherapy: People who received chemotherapy treatment for a previous cancer have a higher chance of developing leukemia later in life.

Exposure to benzene: This is a solvent that manufacturers use in some cleaning chemicals and hair dyes.

Some genetic conditions: Children with Down syndrome have a third copy of chromosome 21. This increases their risk of acute myeloid or acute lymphocytic leukemia to 2–3%, which is higher than in children without this syndrome.

Another genetic condition with links to leukemia is Li-Fraumeni syndrome. This causes a change to the TP53 gene.

Family history: Having siblings with leukemia can lead to a low but significant risk of leukemia. If a person has an identical twin with leukemia, they have a 1 in 5 chance of having the cancer themselves.

Inherited problems with the immune system: Certain inherited immune conditions increase the risk of both severe infections and leukemia. These include:

•ataxia-telangiectasia

•Bloom syndrome

•Schwachman-Diamond syndrome

•Wiskott-Aldrich syndrome

Immune suppression: Childhood leukemia may develop due to the deliberate suppression of the immune system. This might occur following an organ transplant as a child takes medications to prevent their body from rejecting the organ.

Several risk factors need further studies to confirm their link to leukemia, such as:

•exposure to electromagnetic fields

•exposure to certain chemicals in the workplace, such as gasoline, diesel, and pesticides

•smoking

•using hair dyes

Eventually, the cancerous cells outnumber healthy cells in the blood.

Types

 There are four main categories of leukemia:

acute

chronic

lymphocytic

myelogenous

Chronic and acute leukemias

During its lifespan, a white blood cell goes through several stages.

Chronic leukemia progresses more slowly. It allows for the production of more mature, useful cells.

Acute leukemia overcrowds the healthy blood cells more quickly than chronic leukemia.

Lymphocytic and myelogenous leukemias

Lymphocytic leukemia occurs if cancerous changes affect the type of bone marrow that makes lymphocytes. A lymphocyte is a white blood cell that plays a role in the immune system.

Myelogenous leukemia happens when the changes affect bone marrow cells that produce blood cells, rather than the blood cells themselves.

Treatment

The primary treatment for leukemia is chemotherapy. A cancer care team will tailor this to suit the type of leukemia.

If treatment starts early, the chance of a person achieving remission is higher.

Types of treatment include:

Watchful waiting: A doctor may not actively treat slower growing leukemias, such as chronic lymphocytic leukemia (CLL).

Chemotherapy: A doctor administers medications intravenously (IV), using either a drip or a needle. These target and kill cancer cells. However, they can also damage noncancerous cells and cause severe side effects, including hair loss, weight loss, and nausea.

Chemotherapy is the primary treatment for AML. Sometimes, doctors may recommend a bone marrow transplant.

Targeted therapy: This type of treatment uses tyrosine kinase inhibitors that target cancer cells without affecting other cells, reducing the risk of side effects. Examples include imatinib, dasatinib, and nilotinib.

Many people with CML have a gene mutation that responds to imatinib. One study found that people who received treatment with imatinib had a 5-year survival rate of around 90%.

Interferon therapy: This slows and eventually stops the development and spread of leukemia cells. This drug acts in a similar way to substances that the immune system naturally produces. However, it can cause severe side effects.

Radiation therapy: In people with certain types of leukemia, such as ALL, doctors recommend radiation therapy to destroy bone marrow tissue before a transplant.

Surgery: Surgery often involves removing the spleen, but this depends on the type of leukemia a person has.

Stem cell transplantation: In this procedure, a cancer care team destroys the existing bone marrow with chemotherapy, radiation therapy, or both. Then, they infuse new stem cells into the bone marrow to create noncancerous blood cells.

This procedure can be effective in treating CML. Younger people with leukemia are more likely to undergo a successful transplant than older adults.

Symptoms of leukemia include the following:

Poor blood clotting: This can cause a person to bruise or bleed easily and heal slowly. They may also develop petechiae, which are small red and purple spots on the body. These indicate that blood is not clotting properly.

Petechiae develop when immature white blood cells crowd out platelets, which are crucial for blood clotting.

Frequent infections: The white blood cells are crucial for countering infection. If white blood cells are not working correctly, a person may develop frequent infections. The immune system may attack the body’s own cells.

Anemia: As fewer effective red blood cells become available, a person may become anemic. This means that they do not have enough hemoglobin in their blood. Hemoglobin transports iron around the body. A shortage of iron can lead to difficult or labored breathing and pale skin.

Diagnosis

A doctor will carry out a physical examination and ask about personal and family medical history. They will check for signs of anemia and feel for an enlarged liver or spleen.They will also take a blood sample for assessment in the laboratory.If the doctor suspects leukemia, they may suggest a bone marrow test. A surgeon extracts bone marrow from the center of a bone, usually from the hip, using a long, fine needle.This can help them identify the presence and type of leukemia.

Outlook

The outlook for people with leukemia depends on the type.

Progress in medicine means that people can now achieve complete remission through treatment. Remission means that there are no longer signs that cancer is present.

When a person achieves remission, they will still require monitoring and may undergo blood and bone marrow tests. Doctors need to carry out these tests to ensure the cancer has not returned.

The doctor may decide to reduce the frequency of testing if the leukemia does not return over time.