Note on thrombotic thrombocytopenic purpura.

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Description

Blood clots (thrombi) form in small blood vessels throughout the body in thrombotic thrombocytopenic purpura, an uncommon illness. If these clots clog veins and restrict blood flow to organs including the brain, kidneys, and heart, they can cause major medical complications. These neurological disorders (such as personality changes, migraines, confusion, and slurred speech), fever, irregular kidney function, abdominal pain, and cardiac difficulties are all possible complications of these clots. Clots form in people with thrombotic thrombocytopenic purpura even when there is no obvious damage. Platelets, aggregates of cells that circulate in the blood and aid in clotting, help to create blood clots. In people with thrombotic thrombocytopenic purpura, a substantial number of platelets are required to form clots, so there are fewer platelets in the bloodstream. This condition also causes early hemolysis (the breakdown of red blood cells). Red blood cells can break apart as flow squeezes past blockages in blood vessels. Hemolytic anemia is a disorder in which red blood cells are destroyed quicker than the body can replace them.

Symptoms

Small clots that form in the smallest arteries can cause a substantial drop in the quantity of blood platelets (thrombocytopenia), improper destruction of red blood cells (hemolytic anaemia), and problems in the neurological system and other organs. Headaches, mental changes, disorientation, speech abnormalities, minor or partial paralysis (paresis), seizures, or coma are all examples of nervous system disturbances. Fever, proteinuria (the presence of blood plasma proteins in the urine), and hematuria (the presence of a small number of red blood cells in the urine) are all possible symptoms. Affected person exhibit red rash-like areas of skin or patches of purplish discoloration (purpura) resulting from abnormal bleeding into the mucous membranes (the thin, moist layer lining the body's cavities) and into the skin that can be a sign of low platelets. Additional features of TTP can include abnormally heavy bleeding (hemorrhaging), weakness, fatigue, lack of colour (pallor), and abdominal pain with nausea and vomiting. The individuals with TTP, increased levels of a chemical compound known as creatinine are found in the blood.

Cause

The exact cause of TTP is not known. It is associated with a deficiency of an enzyme involved in blood clotting called the von Willebrand factor cleaving protease (also called ADAMTS13). The deficiency of this enzyme allows large complexes of the clotting protein known as von Willebrand factor to circulate in the blood, resulting in platelet clotting and the destruction of red blood cells.

Treatment

Plasmapheresis, also known as plasma exchange, is a procedure that removes antibodies that inhibit the ADAMTS13 protease while also reintroducing the functioning ADAMTS13 protein. During this procedure, a machine removes blood from the affected individual, separates blood cells from plasma, replaces the patient's plasma with healthy plasma, and returns the blood to the patient. Steroids are also administered to patients to stop the formation of anti-ADAMTS13 antibodies. Rituximab, an anti-CD20 antibody, is also extensively used in the treatment of iTTP to decrease the generation of anti-ADAMTS13 antibodies, with a considerably longer impact than steroids.

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