Primary Immune Thrombocytopenia

Primary Immune Thrombocytopenia (ITP) is an autoimmune disease most commonly seen in children following a viral illness or vaccination that causes destruction of platelets. It is thought to be mediated by the production of IgG antiplatelet antibodies against GPIIb/IIIa, an important membrane glycoprotein found on platelet membranes. It is often asymptomatic, but can present with bleeding and petechiae. The diagnosis can be made via isolated thrombocytopenia and/or increased number of megakaryocytes on bone marrow biopsy. This disease can be treated with steroids in acute cases, intravenous immunoglobulin, or various immunosuppressants such as rituximab. Splenectomy may be performed in refractory cases.

PLAY PICMONIC

Characteristics

Following Infection or Vaccination

Virus Bacteria-guy Vaccine-syringe

The acute form of ITP is most commonly seen in otherwise well-appearing and healthy children and adolescents following a viral infection. It has also been documented to occasionally occur following measles, mumps, and rubella (MMR) vaccination.

Antiplatelet Antibodies

Ant-tie-plates Ant-tie-body

GPIIb/IIIa is one of the glycoproteins present on platelet membranes. It is also the receptor on platelet membranes that interacts with von Willebrand Factor (vWF) and aids in platelet activation. The pathogenesis of ITP is thought to be related to an autoimmune response against GPIIb/IIIa mediated by IgG antibodies.

Clinical Features

Often Asymptomatic

Thumbs-up

Patients with primary immune thrombocytopenia are often asymptomatic for extended periods. This disease may be discovered incidentally on routine bloodwork, or may present after an extended period of chronic disease.

Bleeding

Dripping Blood

Patients with ITP who are symptomatic may experience prolonged bleeding. Some examples include gingival bleeding while brushing teeth, frequent or prolonged nosebleeds, and prolonged menstrual bleeding.

Petechiae

Tlki guy

Patients with ITP may develop a petechial rash. Petechiae are pinpoint, round spots that appear on the skin due to bleeding in small capillaries. They typically do not blanch when pressure is applied and are flat to the touch.

Diagnosis

Thrombocytopenia

Trombone-side-toe-peanut

An isolated decrease in platelets, while the white blood cell count and hemoglobin remain relatively within normal limits, is characteristic of this disease. This is in contrast to other hematologic disorders such as hematologic malignancies and microangiopathic hemolytic anemias in which multiple cell lines will be affected in addition to platelets.

Increased Megakaryocytes

Up-arrow Megaphone-carrot

Patients with primary immune thrombocytopenia may present with increased number of megakaryocytes on bone marrow biopsy. Recall that megakaryocytes are the progenitor cell to platelets, and therefore systemic destruction of platelets will result in increased production of new platelets to compensate.

Management

Corticosteroids

Quarter-on-steroids

Corticosteroids are appropriate if only a short period of treatment is required, such as in patients with acute ITP that subsequently resolves. Corticosteroids help manage ITP by suppressing the immune system and therefore suppressing the autoimmune response thought to play a pathogenic role in ITP.

Intravenous Immunoglobulin (IVIG)

Ivy-gold-goblin

Administration of IVIG is sometimes indicated in patients with ITP, especially if they have extremely low platelet counts (<30,000), are experiencing severe bleeding, or are unable to tolerate glucocorticoids. IVIG is sufficiently effective that it can raise the platelet count within 24-48 hours, and this effect typically persists for 2-6 weeks. IVIG acts by interfering with macrophage uptake of autoantibody-coated platelets, effectively acting as a decoy in this way.

Rituximab

Red-tux-mob

Rituximab may be used in the treatment of primary immune thrombocytopenia. This medication is an antibody that binds to CD20, a receptor on B cells. Inhibition of this receptor prevents proliferation of these cells, thus decreasing platelet destruction.

Splenectomy

Chopped-off Spleen and Scalpel

In symptomatic patients with thrombocytopenia that is refractory to other treatment options, splenectomy may be necessary. This is thought to help because the spleen plays a role in the elimination and consumption of platelets, and therefore splenectomy can help mitigate this effect. Following splenectomy, patients should be counseled on receiving vaccinations against encapsulated bacteria.