

Practical Problems in Transfusion Medicine

Transfusion Therapy Case Study 1

Part 3: Diagnosis and Treatment

Since this thrombocytopenia has occurred in a healthy individual who has an otherwise normal complete blood count (CBC) and coagulation studies, one can conclude that she may have idiopathic thrombocytopenic purpura (ITP). Before accepting the diagnosis of ITP, however, it should be determined that the patient has not recently ingested any medication or other substance, such as quinine water, that is associated with immune thrombocytopenia. No other workup, including the performance of a bone marrow examination, or tests to detect platelet-associated IgG or serum antibodies directed against platelet surface glycoproteins, are usually required to make the presumptive diagnosis of ITP. In the absence of significant bleeding, transfusion of platelets is inappropriate, and treatment is directed at the underlying immune abnormality with the use of steroids and other medical interventions.

HELPFUL FACTS

Idiopathic Thrombocytopenic Purpura

Idiopathic Thrombocytopenic Purpura (ITP) denotes an immune-mediated disorder that results in the removal of platelets from the circulation by phagocytic cells in the spleen and elsewhere. It may be caused by autoantibodies directed against the various glycoprotein constituents of the platelet glycolyx. These patients are usually adults who develop a chronic thrombocytopenia that may be resistant to treatment. Cases that occur in patients with systemic lupus erythematosus and other autoimmune diseases are identical to those that occur without other associated disorders. ITP may also result from immune complexes attached to the platelet glycolyx by their Fc fragments. In this case the patients are usually children who often have a recent history of viral illness and tend to have an acute self-limited disease. Adult patients infected with HIV may also have immune thrombocytopenia associated with immune complex formation. ITP does not include those cases of immune thrombocytopenia due to antibodies directed against drugs.

HELPFUL FACTS

Drug-related Thrombocytopenia

The list of drugs associated with immune-mediated thrombocytopenia is lengthy. It was previously thought that most of these drugs caused thrombocytopenia by an immune complex mechanism. In these cases, antibodies bound to drugs dissolved in the plasma and the formed immune complexes were then bound to Fc receptors on circulating platelets. More recently, it has been found that drug-dependent antibodies recognize one or more of the glycoproteins of the platelet glycolyx. The most common drug-related thrombocytopenia among hospitalized patients is heparin-induced thrombocytopenia. In this case, the antibody is directed against heparin that has been complexed to platelet factor 4 released to the platelet surface.

HELPFUL FACTS

Bone Marrow Examination in Thrombocytopenia

The bone marrow of patients with ITP and other immune thrombocytopenias would be expected to show normal to increased numbers of megakaryocytes, while patients with thrombocytopenias not due to peripheral destruction would demonstrate a marked decrease in megakaryocytes. In most cases of ITP, however, the performance of a bone marrow aspirate/biopsy is not essential to make the diagnosis. As in this case, patients with newly diagnosed ITP present with a sudden onset of isolated thrombocytopenia and are in otherwise good health. This strongly suggests that bone marrow failure is almost certainly not the cause of the thrombocytopenia and that peripheral destruction is responsible for the decreased platelet count. Examination of the bone marrow might be reserved to resolve unusual diagnostic dilemmas in which ITP is suspected in a patient with other systemic diseases that may affect bone marrow function.

HELPFUL FACTS

Treatment of ITP

The survival of transfused platelets will be extremely short in the ITP patient, and the patient will also be exposed to the risks of transfusion, including alloimmunization and infectious disease. Transfusion of platelets should be reserved for those patients with immune thrombocytopenias who are experiencing significant bleeding. Treatment with corticosteroids (prednisone 1-2 mg/kg/day in divided doses) forms the basis for front – line management of ITP. Patients considered to be at high risk for significant bleeding, such as those with hemorrhagic mucosal bullae, or patients scheduled to undergo a surgical procedure, should be considered for early treatment with IVIgG or Rh Immune Globulin.

HELPFUL FACTS

Platelet Antibody Tests

While it is true that the platelets of patients with ITP will usually be coated with IgG antibodies that would be detected with a test for platelet-associated IgG, the diagnostic specificity of the test is poor. Platelets from patients with a wide variety of systemic illnesses complicated by severe thrombocytopenia may also be coated with increased amounts of IgG. Such conditions include sepsis, disseminated intravascular coagulation (DIC), and thrombotic thrombocytopenic purpura (TTP). Tests to detect the presence of antibodies in the plasma that are directed against platelet surface glycoproteins are generally not required to make the diagnosis, but they may be helpful to confirm the diagnosis in those cases of suspected ITP that appear to be refractory to first line therapies.