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Acute Immune Thrombocytopenia Purpura

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Introduction

Immune thrombocytopenic purpura (ITP), formerly known as idiopathic thrombocytopenic purpura, is a relatively rare, but potentially life threatening autoimmune disorder that involves the destruction of platelets by autoantibodies. The resultant decreased platelet count leaves the patient at risk for excessive bruising and bleeding (Hunt, 2010). ITP can occur in both adults and children, and also occurs as an acute or chronic disorder. It is thought that the cause of acute ITP may differ from that of chronic ITP, however, the underlying disease processes that occur are very similar (Johnsen, 2012). Acute ITP, the most common form of the disease, most often occurs in children (Hunt, 2010). It is estimated that acute ITP occurs in as many as 6.4 per 100,000 children per year (Deane, 2014). Most children diagnosed with acute ITP have a history of a recent viral infection, usually occurring three to four weeks prior to diagnosis. The majority of cases are relatively mild and nearly 50 percent of these cases will resolve within two months. The most serious, but very rare, complication of ITP is intracranial hemorrhage (Maher, 2014).

Throughout my experience in pediatric hematology/oncology, I have cared for a number of patients diagnosed with acute ITP. Although ITP is generally treated by specialists in hematology, most patients present to their primary care practitioner with the onset of signs and symptoms. Thrombocytopenia can also be seen in other hematological and oncological diseases and often, ITP is a diagnosis of exclusion. It is important that primary care practitioners that treat both adults and children recognize the presenting symptoms of this disease to begin the proper work-up. Although this disorder is rare, it is essential for practitioners to be aware of this condition to consider it as a potential diagnosis and identify patients at risk for severe complications of the disease.

Pathophysiological Processes

Acute ITP is a complex autoimmune disorder characterized by isolated thrombocytopenia. ITP can involve both increased platelet destruction and decreased platelet production (Kime, Klima, Rose & O'Brien, 2013). ITP results from autoantibody immunoglobulin G (IgG) binding to glycoproteins, typically GPIIb/IIIa, on the surface of the platelets (Bredlau, Semple, & Segel, 2011). IgG that is bound to circulating platelets serves as an opsonin (a molecule that enhances phagocytosis by marking an antigen for an immune response) and induces increased platelet phagocytosis and destruction by mononuclear macrophages primarily occurring in the spleen (Lo & Deane, 2014). Anti-platelet antibodies also combine with the antigen to form an immune complex which leads to activation of complement, and results in phagocytosis and increased lysis of platelets (Johnsen, 2012).

Acute ITP is often preceded by a viral infection which is thought to trigger this autoimmune response. The concept of antigenic mimicry may occur in response to this viral infection. The antigen on the viral surface may closely resemble the antigen on the platelet surface leading to the antibodies recognizing the platelets as foreign resulting in platelet removal by phagocytosis (Abadi, Yarchovsky-Dolberg & Ellis, 2015).

Recent research has shown that with ITP, there is also a dysfunction of megakaryocytes (large cells in the bone marrow that break into fragments that become platelets). Megakaryocyte production and differentiation is stimulated by the hormone thrombopoietin. Thrombopoietin is secreted by the liver in response to alterations in platelet count, however, this response is impaired in patients with ITP (Lu & Nossent, 2015). This impaired production of thrombopoietin leads to abnormalities in megakaryocytes and a decreased production of platelets (Labarque & Geet, 2014). The IgG autoantibodies may also bind with proteins on the membrane of megakaryocytes impairing the fragmentation of megakaryocytes into platelets and inducing apoptosis (Maher, 2014).

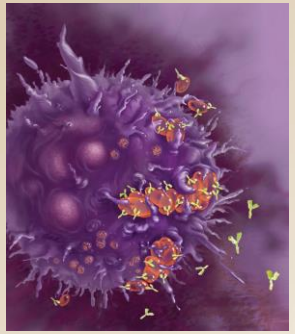

Figure 1. Coating of Platelets with Autoantibodies. This figure shows the binding of IgG to platelet surface. Adapted from "Idiopathic thrombocytic purpura", by D. Winkeljohn, 2010, 14(4), p. 411.

Acute ITP is usually a self-limiting disorder that resolves within six months, but it can progress to the chronic form of the disorder which patients may deal with for a lifetime. While the exact etiology of ITP is unknown, acute ITP is often preceded by a viral infection a few days or weeks prior to the onset of symptoms. The majority of the time, no pathogen is identified but in a small number of cases, Epstein-Barr virus, influenza and varicella-zoster virus have been detected. ITP is also a known, but very rare complication of the measles-mumps-rubella vaccine (Johnsen, 2012). This complication corresponds with the concept of antigenic mimicry or provocation of the immune system resulting in an autoimmune response. Patients with one autoimmune disease are at a higher risk of developing a second autoimmune disease. ITP has been shown to arise in individuals with systemic autoimmune diseases such as systemic lupus erythematous and rheumatoid arthritis (Johnsen, 2012).

ITP occurs with varying levels of thrombocytopenia. Patients that present without any signs of active bleeding and a platelet count greater than 10,000/mm^3 usually do not require treatment (Buchanan, 2014). However, patients with platelet counts less than 10,000/mm^3, active bleeding, comorbidities, or any risk for trauma will be treated with drug therapies (Bredlau, Semple & Segal, 2011).

Case Study

A mother brought her previously healthy two-year-old son to his primary care provider with concerns about sudden onset scattered bruising to his bilateral legs, arms and trunk, purple freckles to his bilateral upper extremities, and a twenty minute nosebleed. The patient's mother denied that her son had experienced any pain, fever, fatigue, recent weight loss, headache, or recent trauma. The patient had a positive history of upper respiratory infection about three weeks ago per the patient's mother. The patient's mother reported no relevant past medical history, no current medication use, and no family history of bleeding disorders or cancers.

On physical exam, the patient was noted to be very active with vital signs within normal limits for the patient's age. Temperature 99.0°F, heart rate 114 beats/minute, blood pressure 92/58, respiratory rate 28 breaths/minute, and oxygen saturation of 99% on room air. No pallor was noted, but significant scattered ecchymosis and petechiae were present to patient's trunk and bilateral upper and lower extremities. The remainder of the physical exam was normal, specifically no lymphadenopathy, splenomegaly, or hepatomegaly were noted (Labarque & Geet, 2014).

A complete blood count (CBC) was obtained and examination of peripheral blood smear was performed (Labarque & Geet, 2014). The patient's CBC showed a normal white blood cell (WBC) count of 6,800/mm^3, normal hemoglobin (Hgb) of 13.7g/dl, and a decreased platelet count of 17,000/mm^3. The peripheral blood smear appeared normal with the exception of an increase in average platelet size (Maher, 2014).

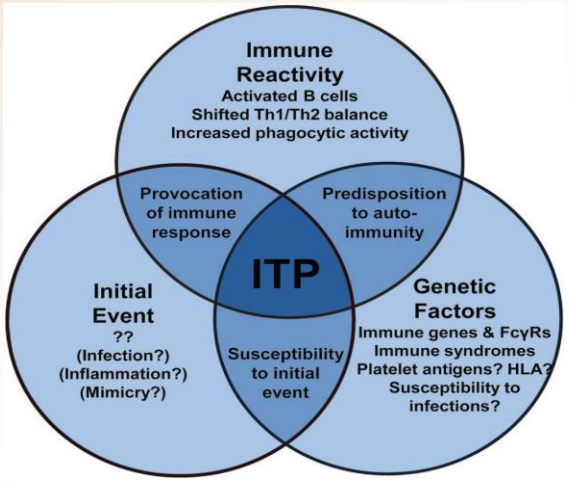

Figure 2. Model of relationship of contributing factors in ITP. Adapted from "Pathogenesis in immune thrombocytopenia : New insights", by J. Johnsen, 2012,. American Society of Hematology, 2012(1), p. 310.

Signs and Symptoms

ITP is often a diagnosis of exclusion and patients can present with no obvious symptoms. ITP is generally defined by a platelet count of less than 100,000/mm^3 (Lo & Deane, 2014). The thrombocytopenia can be an incidental finding on a routine CBC because signs and symptoms associated with ITP may not be apparent until the platelets drop below 30,000/mm^3.

Acute ITP usually has an abrupt onset of symptoms and can occur in otherwise healthy individuals (Maher, 2014). Common signs of ITP are easy bruising, petechiae, prolonged bleeding from cuts or scrapes, epistaxis, menorrhagia, and mucosal bleeding (Hunt, 2010). Blood may be present in the urine, stool, or sputum, but this is much less common (Maher, 2014). Intracranial hemorrhage is the most dangerous, but very rare complication of ITP. Individuals with intracranial hemorrhage may complain of headaches, blurry vision, or display a change in or loss of consciousness. Individuals that present with mucosal bleeding are more likely to experience intracranial hemorrhage (Hunt, 2010).

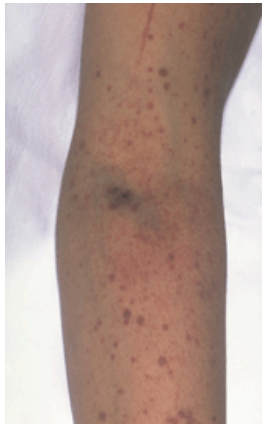

Figure 3. Petechiae and Purpura. Bleeding under the skin causes the red and purple discoloration. Cleveland Clinic Foundation (2012) Retrieved from http://my.clevelandclinic.org

Nursing Implications

Acute ITP is a disorder that is most often managed by specialists in hematology. All nursing professionals should have a general understanding of the disease process because nurses of all levels and specialties may come in contact with this disorder at some time. Patients are likely to present to their primary care provider with the onset of signs and symptoms of ITP. This provider should be able to recognize and identify the potential for this disorder and also have an understanding of the potential complications that may arise as a result of ITP. Having this understanding is essential to performing appropriate testing and making recommendations for further management including possible referral to a specialist. Although the cause of ITP is still under investigation, certain infections and vaccinations do precede some cases. Patients diagnosed with other autoimmune disorders are also more likely to develop ITP throughout their lifetime (Johnsen, 2012). The increased risk for bleeding associated with ITP can also place patients at a much higher risk of developing bleeding complications from medical procedures (Hunt, 2010). To implement appropriate interventions that can prevent complications from these procedures, an understanding of ITP is necessary.

Conclusion

ITP is an autoimmune disorder in which the body attacks its own platelets. ITP occurs in both an acute and chronic form. Acute ITP typically occurs in otherwise healthy children but can also occur in teenagers and adults of all ages. Acute ITP is associated with a sudden onset of symptoms (Maher, 2014). This disorder is associated with an increased risk for bleeding and therefore, increased awareness and recognition of this disorder is key to avoiding the potential complications.

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References

Abadi, U., Yarchovsky-Dolberg, O., & Ellis, M. (2015). Immune thrombocytopenia: Recent progress in pathophysiology and treatment. *Clinical and Applied Thrombosis/Hemostasis, 21*(5), 397-404.

Bredlau, A., Semple, J., & Segel, G. (2011). Management of immune thrombocytopenic purpura in children. *Pediatric Drugs, 13*(4), 213-215.

Buchanan, G. (2014). Immune thrombocytopenia during childhood: New approaches to classification and management. *The Journal of Pediatrics, 165*(3), 437-439.

Hunt, C. (2010). Immune thrombocytopenia purpura. *MedSurg Nursing, 19*(4), 237-239.

Johnsen, J. (2012). Pathogenesis in immune thrombocytopenia: New insights. *American Society of Hematology, 2012*(1), 306-312.

Kime, C., Klima, J., Rose, M., & O'Brien, S. (2013). Patterns of inpatient care for newly diagnosed immune thrombocytopenia in U.S. children's hospitals. *Pediatrics, 131*(5), 880-885.

Labarque, V. & Geet, C. (2014). Clinical practice: Immune thrombocytopenia in paediatrics. *European Journal of Pediatrics, 173*(2), 163-172.

Lo, E. & Deane, S. (2014). Diagnosis and classification of immune-mediated thrombocytopenia. *Autoimmunity Reviews, 13*, 577-583. doi:10.1016/j.autrev.2014.01.026

Lu, C., & Nossent, J. (2015). Thrombopoietin levels in systemic lupus erythematosus are linked to inflammatory cytokines, but unrelated to thrombocytopenia or thrombosis. *Lupus, 24*(1), 18-24. doi:10.1177/0961203314547796

Maher, G. (2014). Immune thrombocytopenia. *South Dakota Medicine: The Journal of the South Dakota State Medical Association, 67*(10), 415-417.

Winkeljohn, D. (2010). Idiopathic thrombocytic purpura. *Clinical Journal of Oncology Nursing, 14*(4), 411-413. doi:10.1188/10.CJON.411-413