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CASE REVIEW: IDIOPATHIC THROMBOCYTOPENIC PURPURA

General Medicine

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ABSTRACT

Immune thrombocytopenic purpura (ITP) is an autoimmune pathology characterized by a low platelet count, purpura, and hemorrhagic episodes caused by antiplatelet autoantibodies. The exclusion typically makes the diagnosis of the known causes of thrombocytopenia. IgG autoantibodies sensitize the circulating platelets. It leads to the accelerated removal of these cells by antigen-presenting cells (macrophages) of the spleen and sometimes the liver or other components of the monocyte-macrophage system. Bone marrow compensates the platelet destruction by increasing platelet production. ITP most often occurs in healthy children and young adults within a few weeks following a viral infection. Certain drugs can also cause immune thrombocytopenia indistinguishable from ITP. Most children have spontaneous remission within a few weeks or months, and splenectomy is rarely needed. However, young adults rarely have spontaneous remissions necessitating splenectomy within the first few months after diagnosis.

KEYWORDS

Idiopathic thrombocytopenic purpura; ITP; Platelets; purpura; Hematology

INTRODUCTION -

Platelets are released from the megakaryocyte, likely under the influence of flow in the capillary sinuses. The normal blood platelet count is 150,000-450,000/μL. The major regulator of platelet production is the hormone thrombopoietin (TPO), which is synthesized in the liver. Synthesis is increased with inflammation and specifically by interleukin 6. TPO binds to its receptor on platelets and megakaryocytes, by which it is removed from the circulation. Thus a reduction in platelet and megakaryocyte mass increases the level of TPO, which then stimulates platelet production. Platelets circulate with an average life span of 7-10 days. Approximately one-third of the platelets reside in the spleen, and this number increases in proportion to splenic size, although the platelet count rarely decreases to <40,000/μL as the spleen enlarges. Platelets are physiologically very active, but are anucleate, and thus have limited capacity to synthesize new proteins.

Thrombocytopenia Results From One Or More Of Three Processes:

(1) decreased bone marrow p production; (2) sequestration, usually in an enlarged spleen; and/or (3) increased platelet destruction. Disorders of production may be either inherited or acquired.

Immune thrombocytopenic purpura (ITP; also termed idiopathic thrombocytopenic purpura) is immune-mediated destruction of is of platelets and possibly inhibition of platelet release from the megakaryocyte. ITP has two types –Acute and chronic. In children, it is usually an acute disease, most commonly following an infection, and with a self-limited course. In adults, it is a may more chronic disease, although in some adults, spontaneous remission most occurs, usually within months of diagnosis. ITP is termed secondary if it is associated with an underlying disorder; autoimmune disorders, on- particularly systemic lupus erythematosus (SLE), and infections, such her as HIV and hepatitis C, are common causes. The association of ITP with Helicobacter pylori infection is unclear but appears to have a geographic distribution.

Clinical Features-

In adult many patients with ITP are diagnosed as a result of chronic asymptomatic thrombocytopenia. For patients with symptoms, primarily related to thrombocytopenia and bleeding, but patients also feel fatigue and decrease quality of life.ITP is characterized by mucocutaneous bleeding ,ecchymoses and petechiae, or with thrombocytopenia incidentally found on a routine CBC. Mucocutaneous bleeding, such as oral mucosa, epistaxis, gastrointestinal, or heavy menstrual bleeding, may be present. Rarely, life-threatening, including central nervous system, bleeding can occur. Wet purpura (blood blisters in the mouth) and retinal hemorrhages may herald life-threatening bleeding.

Diagnosis-

ITP remains as a diagnosis of exclusion that is made in patients with isolated thrombocytopenia. ITP is suspected when there is significant clinical history and physical examination. first line of investigation is complete blood count with peripheral smer suggestive of low platelet count. sometime peripheral smear shows giant platelets with otherwise normal morphology.

Laboratory testing for antibody is usually not recommended because of low sensitivity and specificity. Bone marrow examination is recommended for old age (>60yr) for rule out MDS (myelodysplastic syndrome) and patients who did not responds to initial treatment.In bone marrow biopsy its mainly shows normal or higher megakaryocytes.

Laboratory testing is performed to evaluate secondary cause of thrombocytopenia and should include testing systemic lupus erythematosus(SLE),serum protein electrophoresis ,immunoglobulin level mainly detected hypogamma globulinemia,selective IgA deficiency and infection such as HIV, hepatitis C and coagulation study , H.pylori infection.

Treatment-

The treatment of ITP uses drugs that decrease reticuloendothelial uptake of the antibody-bound platelet, decrease antibody production, and/or increase platelet production. The diagnosis of ITP does not necessarily mean that treatment must be instituted. Patients with platelet counts >30,000/μL appear not to have increased mortality related to the thrombocytopenia.

Initial treatment in patients without significant bleeding symptoms, severe thrombocytopenia (<5000/μL), signs of impending bleeding (such as retinal hemorrhage large oral mucosal hemorrhages) can be instituted as an outpatient using single agent. Traditionally, this has been prednisone at 1 mg/kg, although Rh(D) immune globulin therapy (Win Rho SDF), at 50-75 μg/kg, is also being used in this setting. Rh (D) immune globulin must be used only in Rh-positive patients because the mechanism of action is production of limited hemolysis, with antibody-coated cells "saturating" the Fc receptors, inhibiting Fc receptor function. Monitoring patients for 8 h after infusion is now advised by the FDA because of the rare complication of severe intravascular hemolysis. Intravenous gamma globulin (IV IgG), which is pooled, primarily IgG antibodies, also blocks the Fc receptor system, but appears to work primarily through different mechanism(s). IVIgG has more efficacy than anti-Rh (D) in post splenectomized patients. IVIgG is dosed at 1-2 g/kg total, given over 1-5 days Side effects are usually related to the volume of infusion and infrequently include aseptic meningitis and renal failure. All immunoglobulin preparations are derivedfrom human plasma and undergo treatment for viral inactivation.

For patients with severe ITP and/or symptoms of bleeding, hospital admission and combined-modality therapy is given using high-dose glucocorticoids with IVIgG or anti-Rh,(D) therapy and, as needed, additional immunosuppressive agents. Rituximab, an anti-CD20 (B cell) antibody, has shown efficacy in the treatment of refractory ITP, although long-lasting remission only occurs in -30% of patients.

Splenectomy has been used for treatment of patients who relapse after glucocorticoids are tapered. Splenectomy remains an important treatment option; however, more patients than previously thought will go into a remission over time. Observation, if the platelet count is high enough, or intermittent treatment with anti-Rh (D) or IVIgG, or Cyclosporine, Mycophenolate Mofetil and chemotherapeutic agents such as azathioprine, vinca-alkaloid, cyclophosphamide and interferon have been used with similar success. Vaccination against encapsulated organisms (especially pneumococcus, but also meningococcus and Haemophilus influenzae, depending on patient age and potential exposure) is recommended before splenectomy, Accessory spleen(s) are a very rare cause of relapse.

TPO receptor agonists are available for the treatment of ITP. This approach stems from the finding that many patients with ITP do not have increased TPO levels, as was previously hypothesized. TPO levels reflect megakaryocyte mass, which is usually normal in ITP.TPO levels are not increased in the setting of platelet destruction. Two agents, one administered subcutaneously (romiplostim) and another orally (eltrombopag), are effective in raising platelet counts in patients with ITP and are recommended for adults at risk of bleeding who relapse after splenectomy or who have been unresponsive to at least one therapy, particularly in those who have a contraindication to splenectomy.

We present a case report on ITP. Although not an uncommon entity, there is still much to be learned and this case review will summarize some of the key points in the diagnosis and management of ITP.

Case Report

A 36 year old male presented to the OPD for generalized fatigue and purpura in bilateral upper limb since 2 days .he is apprentaly asymptomatic before that. After further questioning patient admitted to having 1 episode of hematuria which were self-limited. Patient denied epistaxis, hemoptysis, gingival or gastrointestinal bleeding.

The patient have no comorbidity with no significant family history or bleeding disorders were reported. He denied alcohol, drugs or tobacco abuse.

Upon arrival to the opd he was alert, awake and oriented, with a blood pressure of 127/72 mmHg, heart rate 100 bpm, temperature 98.4 $^{\circ}$F, respiratory rate 18 breaths/min. No orthostatic changes were documented. Physical examination revealed a cooperative and well-appearing man, not in any apparent distress. The pupils were equally round and reactive to light and accommodation. The neck was supple, there were no carotid bruits. No signs of gum bleeding were noticed. The chest was clear to auscultation bilaterally. The heart rate was regular. Auscultation revealed a normal S1 and S2. There were no additional sounds. Abdominal examination revealed no hepatosplenomegaly, bowel sounds were present, no abdominal tenderness, guarding or rebound was found. The extremities revealed no clubbing or edema. Examination of the skin was remarkable for purpura in bilateral upper limb which is painless and nonitching. Joints were non-tender and non-erythematous, range of movement was normal. The neurologic examination revealed 5/5 motor responses bilaterally in both upper and lower extremities. No sensory deficits were present. Deep tendon reflexes were 2+ in all four extremities.

Laboratory findings were as follows: Hb 9.8 gr/dL ,Hct 24.4%, WBC: 6.8 x 10^3 per uL, MCV: 84 fL, Platelets: 15,000 per UL, PTT: 28.2 sec, PT: 11.8 sec, INR: 1.02. WBC smear was normal. RBC smear showed hypochromic, macrocytic 1+, microcytic 1+, anisocytosis, basophilic stippling. HIV and Hepatitis – negative.coagulation profile –normal. SLE profile normal. Abdominal ultrasound showed mild splenomegaly with normal liver. serum electrophoresis –normal, Bone marrow biopsy shows increased megakaryocytes. so above history in examination diagnosis of ITP made.

The patient was admitted to the intensive care unit for close monitoring. Injection methy prednisolone 500mg for 3 days given and than Tablet prednisolone 1mg/kg given in tapering dose for 1 month. Platelets improved after injectable prednisolone to 147,000 per UL .than patient discharged with tapering dose of steroid . After 20 days patient again visit to emergency department with bilateral lower limb purpura .he was admitted in ICU and his platelet count at this time was 8000/ul .The patient was started on intravenous immunoglobulin dose of 1-5 gm/kg total for 5 days and Tablet Azathioprine 100mg daily,good response treatment and subsequent rise of platelet count up to 1,50,000/ul. patient is stamp as steroid resitant ITP.