Case reports

Corticosteroid Resistant Idiopathic Thrombocytopenic Purpura: Case Report and Literature Review

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ABSTRACT Idiopathic thrombocytopenia purpura (ITP), also known as immune thrombocytopenic purpura, is defined as isolated thrombocytopenia with normal bone marrow aspirate and the absence of other causes of thrombocytopenia. The first line of treatment of ITP is corticosteroids. We present a case of a patient who was diagnosed with ITP but his ITP was resistant to corticosteroids.

Introduction

The differential diagnoses of a patient with decreased number of platelets:

A. Disorder of platelet production
1. Viral Infections (rubella, mumps, varicella, Hep C, EBV, HIV)
2. Bone marrow disorder (hypoplasia, megakaryocytic aplasia, alcohol induced marrow suppression)

B. Disorders of Platelet destruction
1. Idiopathic thrombocytopenic purpura (ITP)
2. Thrombotic thrombocytopenic purpura (TTP)
3. Disseminated intravascular coagulation (DIC)
4. Pseudo thrombocytopenia
5. Hypersplenism

Here is illustrated a case of decreased platelets fitting with diagnosis of steroid resistant Idiopathic thrombocytopenic purpura (ITP).

Case report

A sixty year old Hispanic male presented with the following complaints with the complaints of flu like symptoms and sore throat from last 2 days, associated with fever and chills from last 2 days. This symptoms complex was associated with development of pin point red spots in different parts of body. He also had one episode of epistaxis. There was no significant history of chest pain and shortness of breath, hemoptysis, hematuria, malena or bleeding per rectum. Also there was no history of blood transfusions, drug abuse or recent vaccinations. His history was significant for heavy drinking and smoking.

A 60 year old Hispanic male, of average build with no obvious distress. His blood pressure at time of presentation was 200/120. He had diffuse petechial hemorrhage scattered over the whole body and bruises on both left and right lower legs. Petechial hemorrhages were also noted on the palate. There was no hepato-spleenomegaly.

His investigation were significant were platelet count of 3, 000, glucose of 160, Mg of 1.6 and Phos of 1.9. CAT scan in ER was performed to rule out any interacerebral bleed due to such a low platelet count, which showed no significant finding.

Patient was started in IV steroids and globulins with the diagnosis of ITP. Anti hypertensive therapy was also started because of the high presenting blood pressure. With this treatment, patient's platelet counts came in normal range. IV globulins were stopped and steroids were tapered off and patient was discharged home with a stable platelet count on oral steroids.

But patient again presented after 8 days with platelet counts of 9,000 and multiple petechie and bruises. Patient was again started on IV steroids and globulins and his platelets count began to increase. At this stage he was labeled "steroid resistant ITP". But patient refused for splenectomy. So he was discharged with stable platelet count on oral steroids.

Just in 6 days, he again presented with platelet count of 4,000. He was again started on IV steroids and globulins to get a stable platelet count to plan for elective splenectomy.

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Discussion

In 1997, American Society of Hematology (ASH) described two criteria's for the diagnosis of ITP;

1. Isolated thrombocytopenia with otherwise normal CBC and peripheral smear.
2. No other condition or factors that may cause thrombocytopenia.

ITP is usually divided into two types,

1. Acute ITP
2. Chronic ITP

Acute ITP	Chronic ITP
- More Prevalent in Late Spring to Winter -Prevalent among children <10 y -Peak incidence age 2-5 years -Equal male: female	-Thrombocytopenia for > 6 months -No seasonal predilection -More prevalent in adolescents and adults -3:1 Female to Male -More likely to have underlying autoimmune disease

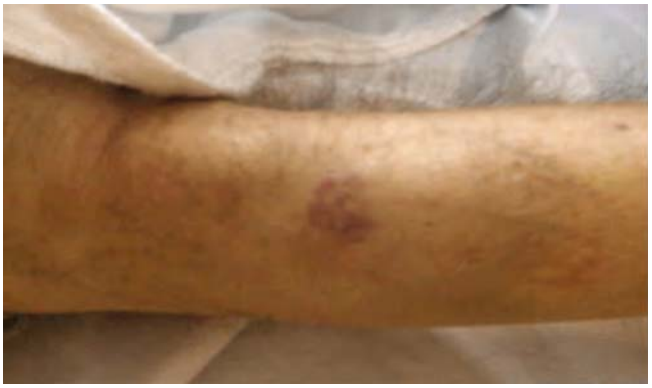

Fig.1 Petechie on legs

Fig.2 Bruises and petechie on leg

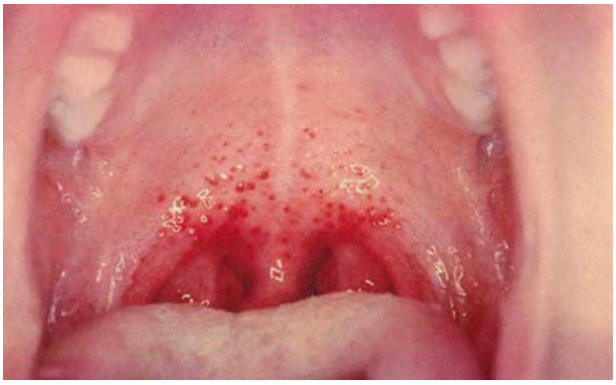

Fig.3 Petechial hemorrhages on the palate

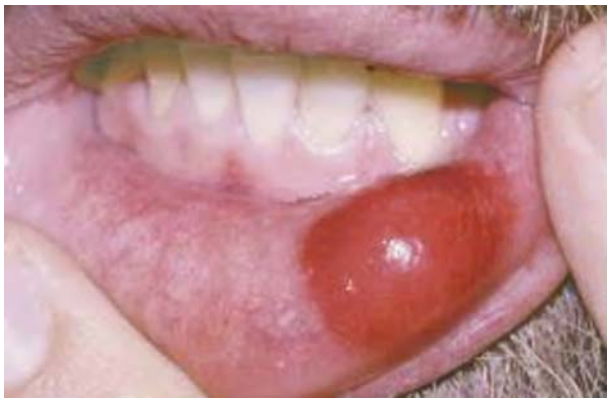

Fig.4 Muco–cutaneous bleeding

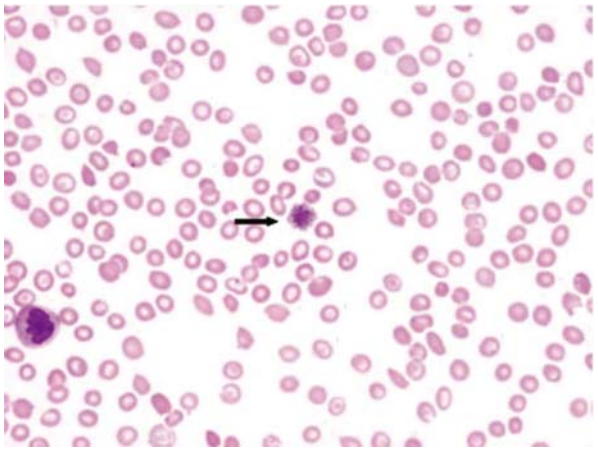

Fig.5 Peripheral smear showing megathrombocytes

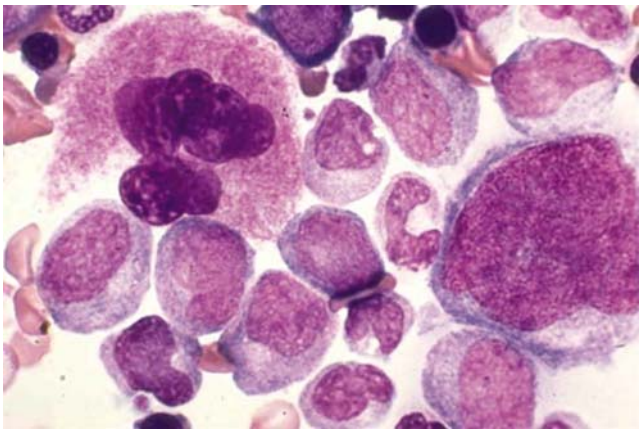

Fig.6 Bone marrow aspirate showing megakaryocytes in the absence of any other abnormality.

ITP is often diagnosed on the basis of routine labs, if thrombocytopenia is mild. Most commonly, patients of ITP usually present with muco-cutaneous bleeding (oral ulcers, epistaxis), menorrhagia, pupura; which is non-palpable, painless and localized to dependent areas.

Less commonly, ITP patients may present as GI bleeding, hematuria, intracranial hemorrhage and hemarthrosis.

Splenomegaly, if present, always suggests an alternate diagnosis.

Laboratory diagnosis

ITP is basically diagnosis of exclusion. On complete blood count, the hall mark of ITP is the isolated thrombocytopenia. On peripheral blood smear, red blood cells and leukocytes are normal. The morphology of platelets is typically normal, with varying numbers of large platelets. Some patients with acute ITP may have megathrombocytes or stress platelets, reflecting the early release of megakaryocytic fragments in to circulation.

Assays for platelet antigen-specific antibodies, platelet-associated immunoglobulin, or other platelet antibodies are available in some medical centers. These tests are usually not recommended by American Society of Hematology because of poor positive/negative predictive values. Testing of anti-platelet antibodies is not required to diagnose ITP (1).

Computed tomography (CT) scanning and magnetic resonance imaging (MRI) are relatively benign and useful non-invasive imaging studies that can be used to rule out other causes of thrombocytopenia.

The primary diagnostic evaluation is bone marrow aspiration and biopsy. Patients with ITP have normal-to-increased number of megakaryocytes in the absence of other significant abnormalities. The cellularity of the aspirate and the morphology of erythroid and myeloid precursors should be normal (2).

Management

Major Bleedings are rare if the platelet counts are more than 10,000. Treatment goal for ITP is to get safe platelet levels and prevent bleeding.

American Society of Hematology gives the following guidelines for the treatment of ITP,

1. Don't treat if,

Platelet count > 50,000 and patient is asymptomatic.

2. Consider treatment if;

Platelet count > 50,000 and there is mucous membrane bleeding.

3. Definitely treat if;

Platelet count < 50,000

4. Hospitalize if;

There is severe bleeding regardless of platelet count or platelet count < 20,000 and mucous membrane are bleeding.

Corticosteroids i.e. oral prednisone, IV methylprednisolone or high-dose dexamethasone is the drug of choice for the initial management of ITP. It's important to get the bone marrow biopsy to confirm the clinical presentation, because corti costeroids may change marrow morphology (3, 4, and 5).

IV immunoglobulin (IVIG) has been the drug of second choice. For Rh (D)-positive patients with ITP and intact spleens, IV Rho immunoglobulin (RHIG) offers comparable efficacy, less toxicity, greater ease of administration and a lower cast (6, 7).

In patients with acute ITP, splenectomy usually results in rapid, complete and life-long clinical remission. In patients with chronic ITP, the results of splenectomy are less predictable.

Refractory ITP

Refractory ITP is defined as platelet count<50,000 for 3 months despite gluco-corticoids and splenectomy. There is no census on when and how to treat this condition. American Society of Hematology makes no recommendations regarding treatment because of poor data quality.

In case of refractory ITP, withhold the treatment if platelet count is 10,000 to 30,000 and there is no significant bleeding. Chronic low-dose corticosteroids, IVIG, Anti-Rho and accessory splenectomy, Danazol and immunosuppressive/chemotherapy agents are the other available options for the refractory ITP.

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