Case Report

ITP- Immune Thrombocytopenia : A Case Report

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Abstract

Until recently, the abbreviation ITP stood for idiopathic thrombocytopenic purpura, but current awareness relating to the immune-mediated nature of the disease, and the absence or minimal signs of bleeding in a large proportion of cases have led to a revision of the terminology. We present a case of Primary immune thrombocytopenia. This article also provides an update on the current definitions related to immune thrombocytopenia, as per recommendations of an International Working Group (IWG) on ITP.

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Introduction

Immune thrombocytopenic purpura (ITP) is an immune mediated acquired disease of adults and children characterized by transient or persistent decrease of the platelet count and depending upon the degree of thrombocytopenia, it is associated with an increased risk of bleeding.¹

Case Report

A 32 year old female patient presented with red to purple discoloured patches over thigh, legs, forearms and back since 6 months. She gave a history of menorrhagia and bleeding gums since 2 months. No h/o fever, headache, epistaxis, haematuria, PR bleeding or malaena. No h/o excessive or delayed bleeding. No family history of similar complaints. The patient had 2 full term normal deliveries which were uneventful.

On examination, the patient was average built, with all vital parameters within normal limits. Multiple red to purple purpuric lesions- mainly petechiae ranging in size from 1-2 mm were seen over thighs, legs, forearm and back. These petechiae were flat, did not blanch on pressure and appeared and regressed over a period of days. (Fig 1) Few ecchymotic lesions were also seen. (Fig 2)The episodes of bleeding were sudden, insidious and intermittent. Examination of the oral cavity revealed inflamed gingiva with bleeding on probing. No lymphadenopathy, hepatomegaly or splenomegaly was noted. No sternal tenderness was present. Ophthalmic examination was normal. No signs of meningeal irritation or raised intracranial tension was noted.

The patient’s haematological workup revealed haemoglobin level of 11.2gm%, total leukocyte count: 11,000/mm³ and differential leukocyte count P₆₀L₃₄E₃M₃. The platelet count was 5,000/mm³ with MPV: 12.0fl and PDW: 12.6fl on admission. The blood counts were done using Sysmex KX-21 and Sysmex XS 800i cell counters. There was marked thrombocytopenia with giant platelets noted on peripheral blood film examination. No platelet satellitism was noted. The tests for haemostasis and blood coagulation- BT, CT, PT, aPTT were normal. The liver function tests and renal function tests were within normal limits. Bone marrow aspiration was performed, which showed increase in the number of megakaryocytes, particularly young hypolobated forms. Serological tests for HIV was non-reactive. Antinuclear antibody was also negative, which ruled out SLE. USG abdomen and CT scan were also normal.

On admission, the patient’s platelet count was 5,000/mm³. She was treated with steroids and given 7 Random donor platelets and one Single donor platelet, which raised the platelet count over a period of 7 days to 1,64,000/mm³. In the next 7 days, the platelet count declined to 11,000/mm³, indicating rapid immune destruction.
**Discussion**

An International Working Group of recognized experts convened in Vicenza, Italy (the Vicenza Consensus Conference) in Oct 2007 and a report was published in 2009 to standardize critical definitions, outcome criteria and terminology related to ITP. 

The panel decided to avoid the term “Idiopathic”, preferring “Immune” to emphasize the immune-mediated mechanism of the disease and to choose “primary” (as opposed to idiopathic) to indicate the absence of any obvious initiating and/or underlying cause. The term “purpura” was felt inappropriate, because bleeding symptoms were absent or minimal in a large proportion of cases. The acronym ITP (now proposed to stand for immune thrombocytopenia) was preserved because of its widespread and time-honoured use.

Primary ITP is an autoimmune disorder characterized by isolated thrombocytopenia (peripheral blood platelet count 1lakh/mm$^3$) in the absence of other causes or disorders that may be associated with thrombocytopenia.

The panel recommended that the term “acute” which has been used to describe a self-limited form of the disease be avoided. In the absence of reliable predictive clinical or laboratory parameters of disease duration, the term “newly diagnosed ITP” was suggested for all cases at diagnosis.

<table>
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<th>Terminology</th>
<th>Current definition</th>
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<tr>
<td>Platelet threshold</td>
<td>&lt;100x10$^9$/L (Previously defined as &lt; 150x10$^9$/L)</td>
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<td>Primary ITP</td>
<td>Absence of secondary causes to account for thrombocytopenia (diagnosis of exclusion)</td>
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<tr>
<td>Secondary ITP</td>
<td>Immune thrombocytopenia due to disease or drug exposure</td>
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<td>Severe ITP</td>
<td>Bleeding needing treatment regardless of platelet count</td>
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<td>Newly diagnosed ITP</td>
<td>From diagnosis to 3 months(previously known as acute ITP until 6 months from diagnosis)</td>
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<td>Persistent ITP</td>
<td>3-12 months after diagnosis</td>
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<tr>
<td>Chronic ITP</td>
<td>&gt;12 months after diagnosis(previously defined as &gt; 6 months after diagnosis)</td>
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ITP has an incidence of up to 6.4 per 100000 children and 3.3 per 100000 adults per year. The presentation and management of ITP are different in adults and children. The peak incidence of ITP in adults is in the age group 15-40 years with a female: male ratio of 1.2-1.7:1. The onset of presentation is insidious. The patients usually present with purpura and most of the symptoms are of greater than 2 months of duration. The platelet count in most cases, is usually <20,000/mm$^3$. Spontaneous remission occurs in less than 2% of cases, and 43% of the patients go in to chronic phase.

Majority of antiplatelet antibodies in patients with ITP are directed against Gp IIb-IIIa and the remainder against the GpIb-IX.complex and other platelet glycoproteins such as Gp IV and Gp Ia- IIa. Most antiplatelet antibodies are IgG. Antibody coated platelets bind antigen presenting cells through FcƳ receptors, primarily in the spleen and in other organs of the mononuclear phagocyte system. Although ITP is antibody mediated, the autoantibodies are under the control of T helper cells and their cytokines. Complement activation plays a role in thrombocytopenia in some patients with ITP.

A significant number of patients are diagnosed incidentally on routine complete blood counts. Symptoms and signs of ITP depend on the platelet count. Approximately one third of patients have platelet counts greater than 30,000/mm$^3$ at diagnosis and no significant bleeding, although bleeding symptoms are generally seen in patients with counts below this level. Purpura (ecchymoses and petechiae), epistaxis, menorrhagia, and gingival bleeding are common. Haematuria, hemoptyisis and gastrointestinal bleeding are less common. Intracerebral haemorrhage is rare. The incidence of life-threatening complications is highest in patients older than 60 years, however mortality rates are low in patients with ITP, even in those with severe thrombocytopenia. The purpuric lesions seen in ITP are not palpable, do not blanch with pressure, and often develop on distal regions of the extremities and on skin exposed to pressure. Haemorrhagic bullae may develop in the buccal mucosa. Bleeding after surgery, trauma, or tooth extraction are common.
Thrombocytopenia is defined as platelet count less than 1 lakh/mm³. Platelet anisocytosis, with increased Mean platelet volume and platelet distribution width is seen in ITP. Platelets may be abnormally large reflecting accelerated production or may be abnormally small reflecting platelet destruction. Autoimmune haemolytic anemia with a positive Coomb’s test and reticulocytosis may accompany ITP, and the association is termed Evan's syndrome. Marrow examination which is not always required to make a diagnosis of ITP, generally reveals normal or increased number of megakaryocytes. American Society of Haematology guidelines for ITP state that marrow aspiration is unnecessary in the evaluation of ITP if the patient is younger than 60 years, has typical presentation, has a good response to first line therapy, and splenectomy is not being considered. Some haematologists recommend that the marrow be evaluated to rule out leukaemia and myelodysplasia, specially in children and those older than 40 years. Phase III assays measure platelet glycoprotein specific autoantibodies. Three techniques that are widely used include immunoblotting, immunoprecipitation, and glycoprotein immobilisation assays. These tests may be useful for discriminating immune from non-immune thrombocytopenia and for monitoring response to treatment. Although these tests are specific, they are not sensitive enough for ITP screening. Treatment of newly diagnosed adult patients or of patients requiring treatment for the first time (initial treatment) is aimed at rapidly obtaining a safe platelet count to prevent or stop haemorrhage and to ensure an acceptable quality of life with minimal treatment related toxicity. The treatment modalities for immune thrombocytopenia range from wait and watch to steroids, immunoglobulins, Anti-D, Rituximab, immunosuppressive agents like Azathioprine and cyclophosphamide, Danazol, Dapsone, splenectomy and Thrombopoietic growth factors. Only in emergency cases, repeated or continuous platelet transfusions may be required. Steroids and or iv anti D or iv IG given prior to transfusions may help preserve platelet longevity in circulation. According to the revised terminology our patient is said to have newly diagnosed ITP. She presented with petechiae and ecchymosis with h/o menorrhagia and bleeding gums. The work up done in the present case revealed thrombocytopenia with raised MPV, normal PDW, normal coagulation profile, bone marrow with increased megakaryocytes. She was treated with steroids and platelets and is on regular follow up.

Conclusion
The diagnosis of primary ITP remains one of exclusion, no robust clinical or laboratory parameters are currently available to establish its diagnosis with accuracy. The main clinical problem of primary ITP is an increased risk of bleeding, although bleeding symptoms may not always be present. All forms of immune-mediated thrombocytopenia except primary ITP should be considered as secondary ITP. It is important to distinguish between primary and secondary immune thrombocytopenia because of their different natural histories and distinct treatments. In case of secondary thrombocytopenia, treatment is often targeted toward the underlying disorder, while drug-induced ITP often remits quickly on withdrawal of the inciting drug.

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References

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