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CASE REPORT

Primary Immune Thrombocytopenia with palpable purpura: a case report.

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Abstract

Immune thrombocytopenia is a relatively common bleeding disorder characterized by isolated destruction of platelets by autoantibodies, with a typically acute onset, benign course and self-limiting in most cases. The aim of this report is to present a rare case of Primary Immune Thrombocytopenia (ITP) with palpable purpura, an uncommon manifestation in this disease and little described in the literature. This is a 12-year-old female adolescent who presented with palpable purpura, ecchymosis and oral cavity bleeding associated with thrombocytopenia and anemia. Initial investigation excluded autoimmune diseases, vasculitis, lymphoproliferative diseases and coagulation disorders, in addition to the myelogram showing typical findings of ITP: hypercellular bone marrow with megakaryocytic hyperplasia and decreased thrombocytogenesis. Due to the diversity of clinical presentation and the possibility of atypical manifestations, the diagnosis is often challenging, as in the case of this patient in which the main finding on physical examination was the presence of palpable purpura.

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INTRODUCTION

Immune thrombocytopenia (ITP) is a relatively common bleeding disorder characterized by isolated destruction of platelets by autoantibodies,[1] typically of acute onset. It is a benign, self-limiting condition in most cases.[2] It manifests in two forms: primary ITP, a condition without a known apparent cause; and secondary ITP, a condition induced by other diseases (autoimmune or lymphoproliferative disorders), drug therapies, or infectious agents.[3]

Primary ITP is more prevalent in the pediatric age group and its incidence is 4.1 to 9.5 per 100,000 children per year, with cases predominating in the age group of one to five years, with a slight predominance in males.[4] It generally occurs after a viral infection or vaccination and is characterized by the formation of autoreactive antibodies, with opsonization of platelets and destruction of the platelet-antibody complex by phagocytes of the reticuloendothelial system mainly in the spleen and liver, leading to thrombocytopenia, the main finding of this condition.[3,5]

ITP is categorized according to the duration of symptoms as acute (up to three months), persistent (from three to 12 months), or chronic (longer than 12 months).[4] In many cases, patients are asymptomatic and diagnosis is made based on an incidental finding of isolated thrombocytopenia in laboratory tests. Clinical manifestations, when present, include predominantly bleeding of the skin, in the form of petechiae and bruises, and of mucous membranes;[3,6] severe hemorrhage and palpable purpura are uncommon.[2]

The diagnosis is based on the exclusion of other causes of thrombocytopenia.[1,7] No specific treatment is generally necessary, as about 80% of the patients progress to spontaneous remission within six months.[5]

This report presents a rare case of ITP with palpable purpura, an atypical manifestation of this disease little described in the literature, and points out the importance of considering non-classic clinical signs of disease in diagnosis. This report was written in compliance with Resolution 466/2012 of the National Health Council and obtained the approval of the Research Ethics Committee of the Professor Fernando Figueira Integral Medicine Institute (certificate no. 57896322.9.0000.5201).

CASE REPORT

A 12-year-old brown girl from the town of Jaboatão dos Guararapes, in the Brazilian Northeastern State of Pernambuco, sought medical care for petechiae and bruises that began to appear three days before admission. They were initially located on her legs and disseminated gradually throughout her body, including the oral cavity. She also had thrombocytopenia (44,000/**µL**) per a test performed at another service. The patient recalled having a similar episode a year ago, with bruises appearing on her neck and arms, which resolved spontaneously, and an isolated episode of epistaxis three months before admission. She did not report spontaneous bleeding, recent infection, or vaccination. During physical examination on admission, the presence of disseminated palpable purpura was observed, predominantly in the arms and legs, along with extensive hematomas (Figure 1).



Figure 1. Palpable purpura, petechiae, and bruises on the legs.

The presence of palpable purpura and the existence of prior bleeding episodes prompted an initial investigation for vasculitis and other autoimmune diseases, in addition to coagulation disorders, all of which were ruled out. Since lab test results indicated she had mild anemia (Hb 10.4) with bicytopenia, a myelogram was ordered to rule out lymphoproliferative diseases. The patient's platelet count dropped gradually to a minimum of 2,000 platelets/µL, without significant involvement of other hematopoietic lineages. The hypothesis of primary immune thrombocytopenia with atypical clinical manifestation was raised. The myelogram results showed a hypercellular bone marrow with megakaryocytic hyperplasia and decreased platelet genesis, with the absence of blast cells in the bone marrow, confirming the diagnosis of ITP.

The significant drop in platelet count and the onset of bleeding in the oral mucosal observed during hospitalization led to the initiation of antifibrinolytic therapy and treatment with corticosteroids, with prednisone administered at a dose of 2 mg/kg/day for five days with a good response. The patient improved, without new episodes of bleeding, and her platelet count increased. She was discharged with 227,000 platelets/mm³ and assigned to outpatient follow-up with the pediatric hematology group.

COMMENTS

ITP is a condition characterized, in most cases, by isolated thrombocytopenia. It includes a wide spectrum of clinical presentations and affects otherwise healthy children without associated systemic symptoms.[1,8] The patient in this report was outside the age group with the highest incidence and was a female. According to the literature, preschoolers are the most affected age group and males are preferentially

involved. However, male and female teens are equally affected.[4]

In most studies, acute ITP is preceded by infection, most often viral, or vaccination. A retrospective study carried out at a tertiary hospital, in which patients were followed up for ten years, found that 35% of patients had a history of disease, 18% with upper airway infections, 15% with acute gastroenteritis, and 3.2% with chickenpox.[9] Our patient had no history of disease.

A study published in 2009 comprising cases of ITP treated at a Brazilian hospital within 12 years showed that hemorrhagic manifestations were seen in 94.7% of the patients, including cutaneous presentations (petechiae and bruises), with fewer cases of bleeding in mucous membranes (epistaxis and gingival bleeding), the gastrointestinal tract, and the genitourinary tract.[10,8]

Despotovic and Grimes (2018) cited the Intercontinental Childhood ITP Study Group cohort, in which hemorrhagic symptoms were reported in 91% of the children at the time of diagnosis.[11] Our patient had thrombocytopenia and mucocutaneous presentations, including petechiae and bruises. Palpable purpura on the legs is a less common finding, which caused the consideration of other conditions in diagnosis.

Purpura is a skin lesion with a red-purple color, which may be macular or elevated. It does not disappear under digital pressure. Palpable purpura is linked to vascular alterations (damage to vessel walls or increased capillary permeability), which may be caused by trauma, infection, vasculitis, or collagen disorders.[12] In ITP, in turn, there is immune-mediated platelet destruction, leading to opsonization and destruction of platelets, with clinical skin manifestations occurring as petechiae and bruises (non-palpable purpura).[13]

Due to the diversity of clinical presentations and the possibility of atypical manifestations, diagnosis is often challenging. In our patient, the main finding on physical examination was palpable purpura. Arnold et al (2017) described the lack of specific criteria as a factor that leads to errors in diagnosis and prescription of inadequate treatment. The authors found that one in seven individuals with the disease was misdiagnosed at some point.[14]

According to the guidelines from the American Society of Hematology, bone marrow testing is not routinely indicated for patients with clinical and laboratory test changes consistent with typical ITP. However, in case of doubt, testing is recommended.[6,15] Our patient presented an unusual manifestation, in addition to anemia concomitant with thrombocytopenia. Therefore, we decided to expand the investigation to improve diagnostic accuracy. The myelogram results were indicative of ITP.

The guidelines recommend expectant management for asymptomatic patients or patients with skin bleeding alone. The platelet count must be followed closely and patients kept on surveillance. Eighty percent of the individuals with the disease improve spontaneously within six months.[13] Treatment is meant to prevent life-threatening hemorrhage and is indicated for children with bleeding in mucous membranes or other sites, with corticosteroids prescribed as first-line treatment.[6,15] Our patient's symptoms guided the choice of therapy. She responded well and was discharged with a normal platelet count.

The rare association of ITP and palpable purpura and the investigation undertaken for our patient, including the diagnostic conclusion, shows the importance of reporting typical pediatric diseases manifesting atypically, a combination that often proves challenging in medical practice.

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