



Case Report

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Thrombotic Thrombocytopenic Purpura: Illustrative Case

Lievi Correa Negrete¹, Alberto Sanjuanelo Fontalvo², Camilo Pelaez Ramos³, Ariel Bello Espinosa^{2,3}, José Bohórquez Rivero^{4*}, Paula Parra Sanchez⁴, Carolina Behaine Bravo⁴ and German Brunal Ramirez⁵

¹Physician, Department of Hematology, IMAT Oncomedica, Colombia

²Physician, Department of Internal Medicine, University of Cartagena, Colombia

³Physician, Internal Medicine, University Hospital of the Caribbean, Colombia

⁴Physician, University of Sinú, Colombia

⁵Physician, University of Cartagena, Colombia

*Corresponding author: José Bohórquez-Rivero, University of Sinu-Cartagena Sectional.

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Abstract

Thrombotic thrombocytopenic purpura is a disease that belongs to the group of thrombotic microangiopathies together with hemolytic uremic syndrome, atypical hemolytic uremic syndrome and microangiopathies of pregnancy. This pathology is characterized by the occurrence of microangiopathic hemolytic anemia, severe thrombocytopenia and organ ischemia related to disseminated platelet-rich microvascular thrombi. It is mainly caused by deficiency or malfunction of the ADAMTS13 protein (a disintegrin and metalloprotease). We illustrate a case of thrombotic thrombocytopenic purpura with a brief review of the literature.

Keywords: Thrombotic thrombocytopenic purpura, Microangiopathic, Anemia hemolytic anemia, ADAMTS13 protein (MeSH)

Introduction

Thrombotic Thrombocytopenic Purpura (TTP) was first described by the physician Eli Moschcowitz in 1921. It is a disease that is part of the Thrombotic Microangiopathies (TMA) along with hemolytic uremic syndrome, atypical hemolytic uremic syndrome and microangiopathies of pregnancy. TTP is characterized by occurrence of Microangiopathic Hemolytic Anemia (MAHA), severe thrombocytopenia and organ ischemia related to disseminated platelet-rich microvascular thrombi. It is primarily caused by deficiency or malfunction of the ADAMTS13 protein (a disintegrin and metalloprotease). The TTP has two spectrums of presentation, it can be acquired due to inhibition by autoantibodies or inherited due to inherited mutations in ADAMTS13.

It is a rare hematologic entity. The incidence of acquired TTP is approximately 3 cases per million adults per year, according to data from the Oklahoma TTP-HUS Registry. Acquired TTP is very rare in children under 18 years of age and is estimated to present in approximately 1 out of 10 million per year. Therefore, in children, the possibility of hereditary rather than acquired TTP should be considered, in contrast to adults, where acquired TTP is approximately 30 times more common than the hereditary TTP [1-3]. At the level of Colombia, there is no information available regarding its epidemiology, the only data on the disease are obtained through case reports [4,5]. We communicate a case of this interesting pathology.



Presentation of the Case

A 44-year-old male with a history of non-Hodking's lymphoma treated in 2012 in remission, presented to the emergency department with a clinical picture of 7 days of evolution characterized by pulsatile headache of insidious onset, frontotemporal location, 6/10 on the analogue pain scale, asthenia, adynamia and paresthesia in the lower limbs. Physical examination evidenced Systolic Blood Pressure (SBP) of 142mmHg, Diastolic Arterial Pressure (DBP) of 89mmHg, Mean Arterial Pressure (MAP) of 102mmHg, Heart Rate (HR) of 106 beats/minute, Respiratory Rate (RF) of 17 breaths/minute, oxygen saturation (SaO₂) of 98% and Temperature (T) of 37.5°C. The patient was conscious, oriented in time, space, and person, with Glasgow 15/15, with no findings of purpura or petechiae. In the admission paraclinical findings, the hemoglobin of 7.5g/dL of macrocytic, hypochromic characteristic was striking (reticulocytes 11.4%, reticulocyte production index of 2), platelets at 27,000 per mm³, hyperbilirubinemia at the expense of indirect (BT:3.0mg/dL, BI:2.5mg/dL, BD:0.5mg/dL), elevated lactate dehydrogenase (1139IU/L), with negative direct antiglobulin test, serum creatinine of 0.8mg/dL and INR at 1. The peripheral blood smear showed macrocytosis, marked polychromatophilia, spherocytes, basophilic stippling, presence of schistocytes in more than 5%, white series with neutrophil predominance without morphological alterations and severe thrombocytopenia (Figure 1). The Elisa test for HIV third generation was negative.

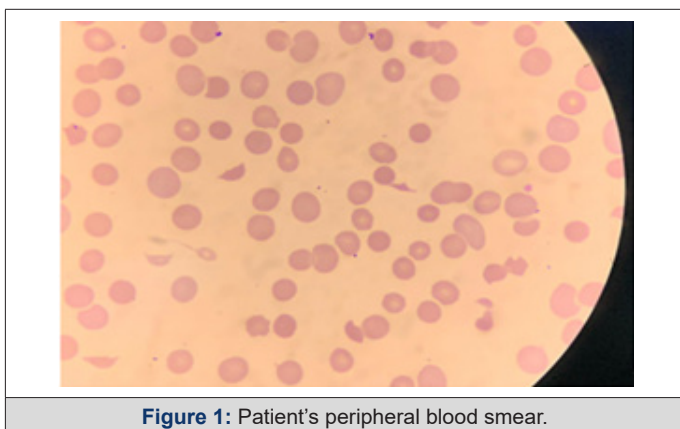


Figure 1: Patient's peripheral blood smear.

These findings were compatible with MAHA with a PLASMIC score of 6 points for a high probability of suffering from a deficiency of ADAMTS13. In view of this, plasmapheresis was initiated urgently, and glucocorticoid methylprednisolone was initiated, with a torpid evolution due to the development of atrial fibrillation, altered state of consciousness, pneumonia associated with health care with the need for prolonged mechanical ventilation and the need for tracheostomy. The diagnosis was confirmed days after initiation of treatment, with an ADAMTS13 activity of 0.2%. The patient was finally discharged from home hospitalization care.

Discussion

The diagnosis of this pathology should be suspected when MAHA (hemoglobin almost always <10 g/dL) and severe thrombocytopenia (platelet count almost always <30,000/microliter) are

present, with or without symptoms of organ affectation and without other clinically apparent etiology [6]. At the time of presumed diagnosis, the PLASMIC SCORE is important because the measurement of ADAMTS13 is not always available in many hospitals, it depends on the background in which the patient is found.

The PLASMIC SCORE was designed to predict the pre-test probability of the activity of ADAMTS13 $\leq 10\%$, it consists of 7 items (see Tables 1 and 2), the higher the score, the greater the possibility of presenting deficiency of that enzyme. The studies suggest that all patients with a PLASMIC score of 5 or more should be treated empirically for TTP unless there is an obvious alternative explanation for the individual's clinical presentation. The diagnosis is confirmed by measurement of ADAMTS13, which should be less than 10%, however levels of $\geq 10\%$ do not discard the possibility of TTP in individuals with a high suspicion for the diagnosis [1,6-9] (Tables 1,2). With respect to treatment, it should be initiated urgently, where the first line will be plasmapheresis and glucocorticoids. In refractory cases, the use of rituximab is indicated [6,10-12].

Table 1: Plasmic score. Taken from Sousa, *et al.*, [12].

Plasmic Score	Points
Platelet count <30,000/microlL	1
One or more indicators of hemolysis: 1. Reticulocyte count >2,5% 2. Undetectable haptoglobin 3. Indirect bilirubin >2.0 mg/dL	1
No active cancer in the previous year	
No history of solid organ transplant or hematopoietic stem cell transplant	1
Mean corpuscular volume <90 femtoliters	1
(INR) <1,5	1
Creatinine <2,0 mg/Dl	1

Table 2: Risk of severe ADAMTS13 deficiency according to PLASMIC SCORE, taken from [8].

Plasmic Score	Risk of Severe Deficiency of ADAMTS13
0-4	Low risk
5	Intermediate risk
7-Jun	High risk

Declaration of Funding Sources

The authors declare that this research was conducted with the authors' own support.

Ethical Considerations

For the realization of this case, informed consent was obtained from the patient and approval was obtained from the institution's ethics committee.

Acknowledgments

None.

Conflicts of Interest

None.

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