Case Report

Cardiology & Research

Purple Thrombocytopenic Thrombotic

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Abstract

Thrombotic thrombocytopenic purpura is a rare disease of immune origin, belonging to thrombotic microangiopathies. In most cases, it can present as microangiopathic hemolytic anemia accompanied by thrombocytopenia, neurological deficit and kidney abnormalities. The present clinical case belongs to a female patient with no significant personal or family history, who went to the doctor for a clinical picture of 2 days of evolution with hematuria, general malaise, asthenia and adynamia, on physical examination without alterations, with vital signs in ranges of normality. The diagnosis of thrombotic thrombocytopenic purpura was made due to hemolysis, elevated levels of lactate dehydrogenase and a reduction in serum haptoglobin accompanied by the presence of serum schistocytes> 6% in the peripheral blood smear. Thrombocytopenic purpura is a diagnostic challenge because its clinical picture is often nonspecific, making it more difficult to start its treatment in a timely manner. Another drawback is the high costs for ADAMTS13 activity tests. Despite the fact that the treatments for this disease have low success rates, in our clinical case our patient responded favorably to the treatment instituted

Keywords:

Thrombocytopenic purpura, thrombotic microangiopathy, treatment

Introduction

Thrombotic thrombocytopenic purpura (TTP) first described by Moschowitz in 1924, which is why it is also called Moschcowitz disease. It is a pathology that is part of the group of thrombotic microangiopathies together with atypical hemolytic uremic syndrome, hemolytic uremic syndrome and microangiopathies of pregnancy (1).

Although it is a fairly infrequent disease, it has an incidence of 4 to 13 cases per million inhabitants per year; It affects women more frequently in a 2: 1 ratio compared to men and its mortality can even reach 90% if it is not treated on time and properly, however, this figure has currently been reduced by around 25% of cases thanks to existing medical therapies (2). It is estimated that 80% of the cases are idiopathic and the remaining 20% are attributed mainly related to viral diseases, autoimmune diseases, immunosuppression states and the use of some medications, in the same way there are reports of multiple presentations of this same disease in individuals of the same family, for which one could speak of a hereditary factor, but there are only around 50 cases reported in the international literatura (3).

Presentation of the case

A 16-year-old female patient with a clinical picture of 3 days of evolution consisting of hematuria, with no significant pathological or family history. Upon admission to the physical examination, the patient was in regular general condition, with vital signs in the normal range, with laboratory tests that showed normochromic normocytic anemia and severe thrombocytopenia with 20,000 platelets in the hemogram, peripheral blood smear with poikilocytosis, schistocytes> 6%, Direct negative coombs, normal urinalysis, LDH in 2000 iu / l, preserved renal function, also with decreased haptoglobin with non-reactive hepatitis B serology.

Herpes 1 and 2 negative, sarscov2 antigen negative, dengue negative, HIV negative. (Table 1). Based on the presence of hemolytic anemia, thrombocytopenia, and elevated LDH, the diagnosis of microangiopathic anemia derived from thrombotic thrombocytopenic purpura was made. Corticosteroid treatment was started, specifically methylprednisolone at a dose of 1 g / day x 3 days, followed by a maintenance dose with prednisone at a dose of 1 mg / kg / day. Due to the poor response to treatment on the second day, plasmapheresis was started.

A sample was sent to perform ADAMTS 13 activity levels, the measurement of which was reported below 4%, which confirmed the diagnosis of thrombotic thrombocytopenic purpura.

Figure 1. Paraclinical report

Paraclinical	Resultado
Hemoglobin	8.1 G/DL
VCM	88 FL
Platelets	20.000
Peripheral blood smear	Poikilocytosis Schistocytes> 6%
Direct coombs	Negative
Uroanalysis	Normal
LDH	2000 UI
BUN	12 MG/DL
Creatinine	0.6 MG/DL
Hepatitis B	Negative
Herpes 1 and 2	Negative
Sars-Cov-2 antigen	Negative
Dengue	Negative
Elisa VIH	Negative
Haptoglobin	Diminished

Discussion

Thrombotic thrombocytopenia purpura, being an atypical presentation pathology, represents a great challenge for its diagnosis because its clinical picture does not always debut clearly, with predominance of microangiopathic hemolytic anemia, fever, thrombocytopenia, renal involvement and neurological abnormalities. what makes it more complicated to start a timely treatment (4). On the other hand, there are several drawbacks with the high costs of ADAMTS13 activity tests, making it difficult to conclude with the diagnosis of this entity. As a differential diagnosis of this pathology in our environment, we must take into account tropical diseases such as leptospirosis, which has similar symptoms. The case is presented due to the rarity of the pathology, highlighting that the patient presented all the clinical characteristics of the disease and the criteria for diagnosis, however, the underlying cause of this could not be established (5).

The follow-up of this disease is carried out with the serial measurement of both platelet levels and LDH levels, since these are the most sensitive indices in monitoring the response to treatment. Treatment with plasmapheresis using fresh frozen plasma, it is important to take it into account in this disease early, because through this the best range of remission and survival is achieved in patients. The current recommendation is to start them as soon as suspected the diagnosis, at a rate of between 1 to 3 volumes per day. Needing in this way between 7 and 16 days to produce a stable remission. The decision to end treatment is empirical and must be estimated based on the clinical and laboratory response, with the exception of renal function values, which may remain permanently altered (6)

To conclude, it can be said that thrombotic thrombocytopenic purpura, despite being a disease with a low frequency, tends to present a high lethality if an early diagnosis and timely treatment is not carried out. However, on the other hand, plasmapheresis has managed to significantly reduce this mortality, for which the recommendation is that in the presence of a patient with the characteristic symptoms of thrombotic thrombocytopenic purpura, a test for ADAMTS13 if possible and start treatment with plasmapharesis (7).

References

1. Hatzistilianou, M., Janoudaki, M., Massaad, M., Geha, R., & Kanariou, M. (2019). Idiopathic Thrombocytopenic Purpura. In Pediatric Immunology (pp. 717-720). Springer, Cham.
2. Oved, J. H., Lee, C. S., & Bussel, J. B. (2017). Treatment of children with persistent and chronic idiopathic thrombocytopenic purpura: 4 infusions of rituximab and three 4-day cycles of dexamethasone. The Journal of pediatrics, 191, 225-231
3. Chandan, J. S., Thomas, T., Lee, S., Marshall, T., Willis, B., Nirantharakumar, K., & Gill, P. (2018). The association between idiopathic thrombocytopenic purpura and cardiovascular disease: a retrospective cohort study. Journal of Thrombosis and Haemostasis, 16(3), 474-480.
4. Ou, Y., Yang, Y., Xiang, X., & Wu, Y. (2020). Relationship between the IL-10 (− 1082 A/G) polymorphism and the risk of immune/idiopathic thrombocytopenic purpura: A meta-analysis. Cytokine, 125, 154820.
5. Zhao, X., Qi, X., Wang, C., Zhou, Z., Cao, H., Wu, P., ... & Wang, Z. (2017). Idiopathic thrombocytopenic purpura: pathogenesis and potential therapeutic approach. Minerva médica, 108(6), 502-506.
6. Hatzistilianou, M., Janoudaki, M., Massaad, M., Geha, R., & Kanariou, M. (2019). Idiopathic Thrombocytopenic Purpura. In Pediatric Immunology (pp. 717-720). Springer, Cham.
7. Takase, K., Kada, A., Iwasaki, H., Yoshida, I., Sawamura, M., Yoshio, N., ... & Nagai, H. (2018). High-Dose dexamethasone therapy as the initial treatment for idiopathic thrombocytopenic purpura: protocol for a multicenter, open-label, single arm trial. Acta Medica Okayama, 72(2), 197-201.