

Diagnosis of thrombotic thrombocytopenic purpura in low income country applicability of plasmic score.

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About the Study

As a medical student, one of the most surprising encounters I had was with a 24-year-old pregnant woman who entered the obstetric emergency department with seizures and cytopenias in 2011. Initially the diagnosis was eclampsia, but soon the name thrombotic thrombocytopenic purpura (TTP) emerged and when plasmapheresis was performed, the patient improved and was able to be discharged and take care of her son. Since then, this disease has gained much of my dedication.

The study "Plasmic score applicability for the diagnosis of thrombotic microangiopathy associated with ADAMTS13-acquired deficiency in a developing country" is a method to answer questions from 2011. Based on the pivotal validation work of the PLASMIC score [1], we tried to determine if the patients treated in our hospital had a correlation between ADAMTS13 activity levels and the clinical manifestations evaluated by those score. The analysis of ADAMTS13 activity was made by ELISA test that was collected in samples before the plasma exchange.

We found that in our population 100% (8 patients) with score above 6 had metalloproteinase activity below 10%. Comparison between the population with PLASMIC score above 6 and below 6 had important differences; the most important were median age (22 vs. 39.5), median days of plasma exchange (6.5 vs. 15.5), pregnancy (100% vs. 0%) and acute kidney injury (100% vs. 0%). In 50% of patients with PLASMIC score above 6 were finding ADAMTS13 inhibitors.

In addition we found that pregnancy, the need for early renal replacement therapy with dialysis and creatinine level above 2 mg/dL were independent factors associated with high levels of ADAMTS13 activity (p-value<0.05). Patients without acquired deficiency of von Willebrand multimer cleavage metalloproteinase, more than half (75%) had an unfavorable outcome, remaining on dialysis after discharge or progressing to death. There is limitation in generalization our findings because the small number of patients.

The evolution of our understanding of TTP has been a milestone for Medicine, making it one of the most fatal diseases in a condition that, but when quickly diagnosed, has a high ability to be controlled. Use of the PLASMIC score is vital and important, especially in developing countries, because of less access to specific tests. In this regard, the present study demonstrated that in our population the PLASMIC score can accurately differentiate patients with TTP and non-TTP.

It is also very important to highlight the fact that currently patients who do not have ADAMTS13 deficiency manifest worse outcome because this group has several conditions, especially complement system amplifying conditions [2], such as hemolytic-uremic syndrome and anti-phospholipid

syndrome. Further studies are needed to assess the applicability of PLASMIC in other locations worldwide as well as to optimize the score depending on the patient group, as the study by Tang et al. [3] has shown.

The current treatment of TTP is based in plasma exchange by apheresis procedures and immunosuppression, such as corticosteroids and monoclonal antibody against B-cell specific mature antigen, rituximab. More recently, caplacizumab had been included on the armamentarium against TTP, specially decreasing the time of thrombocytopenia and the risk of relapse. In low income countries there are some issues for the evidence-based treatment of these patients, especially misdiagnosis of another cause of thrombotic microangiopathy, lack of access to hospitals that can treat these patients with plasma-exchange procedures, lack of access to blood medicine physicians and difficulties to access drugs such rituximab and, more specically, caplacizumab.

We exposed the importance of PLASMIC score to diagnosis of thrombocytopenic thrombotic purpura and the issues related to others diseases, such hemolytic-uremic syndrome and pregnancy related conditions, such pre-eclampsia, eclampsia and HELLP syndrome. It's important note that PLASMIC score is more effective than the former pentad of Moschcowitz to diagnosis TTP, because only 5%-30% of TTP patients have the five points. In our study, there was an undisconsiderable frequency of thrombotic microangiopathy associated to pregnancy (50% of all ADAMTS13 evaluated patients). All the pregnant patients developed acute renal failure, one of them responded to anti-complement treatment. In the present study, all pregnant patients developed the thrombotic microangiopathy after 36 weeks of gestational time and delivery. There is some issues about the importance of complement activation in those context, we strongly recommend that those patients must be evaluated to exclude complement associated thrombotic microangiopathy, specially to the worse prognosis that those diseases have.

References

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