Differentiating TTP from Hypertensive Hemolytic Anemia

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Differentiating Hypertensive Hemolytic Anemia from Thrombotic Thrombocytopenic Purpura

Chelsea Dixon-Dione, D.O., Robert Curtis, D.O., Adele Amine, D.O., Joseph Abbo, M.D., Eva Saha, M.D., Rajlka Munasinghe, M.D.

Introduction

It is established that Focal Segmental Glomerulosclerosis (FSGS) can lead to difficult to control hypertension. It has also been shown that malignant hypertension can cause microangiopathic hemolytic anemia (MAHA), Distinguishing MAHA due to malignant hypertension from other causes of MAHA such as Thrombotic Thrombocytopenic Purpura (TTP) or Hemolytic Uremic Syndrome (HUS) can be difficult. We present an interesting case of a young patient who we believe developed undiagnosed FSGS that led to hypertensive crisis, acute renal failure, and MAHA.

Case Description

Our patient is a 19 year old male with no past medical history who presented to the emergency department with nausea, vomiting, subjective fever, and fatigue for roughly two weeks. He stated that he had been sleeping most of the day, had been unable to keep anything down, and was unable to attend work. He described feeling feverish at some point, but did not take his temperature. Social history was positive for social marijuana use (last use was >1 week prior), social alcohol use. He works part time in the warehouse of a grocery store. Family history was remarkable for early death in his mother, in her mid-30's. Cause of death was unknown, possibly related to an overdose. The patient has been under the care of his aunt ever since.

He had never had surgery, no allergies, and took no daily medications prior to arrival.

Vitals: BP 229/142, HR 92, RR 18, 100% RA, T 98.9°F

The patient was markedly hypertensive on presentation. Physical exam was significant for scleral icterus and the patient was slow to respond to questions, with an otherwise intact neurological exam. Initial blood work revealed renal failure, anemia, and thrombocytopenia. Examination of the peripheral smear demonstrated marked schistocytes.

LDH 1,093
AST 48
ALT 25
T bili 2.1
D bili 0.4
Haptoglobin <30.0

PLASMIC score

<table>
<thead>
<tr>
<th>Score (points)</th>
<th>Risk of Severe ADAMTS13 deficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-4</td>
<td>Low</td>
</tr>
<tr>
<td>5</td>
<td>Intermediate</td>
</tr>
<tr>
<td>6-7</td>
<td>High</td>
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Microangiopathic Hemolytic Anemia (MAHA) - descriptive term for non-immune hemolysis, red blood cell fragmentation that takes place inside the vasculature.

Thrombotic Microangiopathy (TMA) - microvascular thrombosis caused by abnormalities in the vessel walls of capillaries and arterioles. Diagnosis made by tissue biopsy.

FSGS
HTN
MAHA

Microangiopathic Hemolytic Anemia (MAHA) -

PLASMIC Score

Microangiopathic Hemolytic Anemia (MAHA) -

PLASMIC score (points)

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A high PLASMIC score (6.7) has a sensitivity ~91%. A low (0-4) score has a specificity of 99%. The PLASMIC score was applied to 108 patients with suspected TMA. The system had a positive predictive value of 72% and a negative predictive value of 99%. Our patient’s PLASMIC score was 5.

Discussion

This patient’s presentation was concerning for TTP due to severe thrombocytopenia and MAHA. Given the prolonged turnaround time for the ADAMTS13 assay, he was empirically treated for TTP with PEX. However, in hindsight we believe this patient developed FSGS with subsequent renal hypertension that went undiagnosed and untreated for an unknown period of time, leading to his presentation of MAHA secondary to hypertensive crisis.

It is difficult to discern TTP from other causes of MAHA. One helpful tool is called the PLASMIC score which estimates the likelihood of severe ADAMTS13 deficiency in adults with possible TTP.

The average turnaround time for ADAMTS13 is prolonged. Our patient’s assay did not result for >2 weeks. A study at Brigham and Women’s Hospital in 2016 with rapid in-house assay reduced number of plasma exchanges for suspected TTP by more than half without an increase in mortality. A second study in 2017 demonstrated that a rapid assay for ADAMTS13 was associated with reduced mortality for patients diagnosed with TTP.

An article published in 2015 analyzed 19 cases of hypertension induced MAHA via literature review. About 1/3 of the patients underwent PLEX while awaiting ADAMTS13 results. At the time of presentation, all had significant hypertension with MAPs ranging from 123-190 and prominent renal dysfunction, with milder thrombocytopenia.

As the mortality rate of Thrombotic Thrombocytopenic Purpura approaches 90% when not treated with plasma exchange, it is understandable that providers err on the side of caution when choosing whom to treat. Until there is a readily available rapid in-house assay for ADAMTS13, the decision to initiate empiric treatment for TTP will rely on clinical judgment with perhaps some assistance from the PLASMIC scoring system.

References