MAHA ha-ha-ha, get it?

Bryan Takisaki, DO

Providence Health & Services - Spokane Internal Medicine Residency

Introduction

The workup of normocytic anemia is complicated by a broad array of possible etiologies to be considered, each with varying clinical implications.

Microangiopathic hemolytic anemia (MAHA) is an important etiology to be discussed and investigated if indicated by a patient's presentation. The dendritic pathway of investigation is further complicated by understanding the possible causes of MAHA. This careful determination is necessary as it carries significant implications for the treatment and resolution of this anemia.

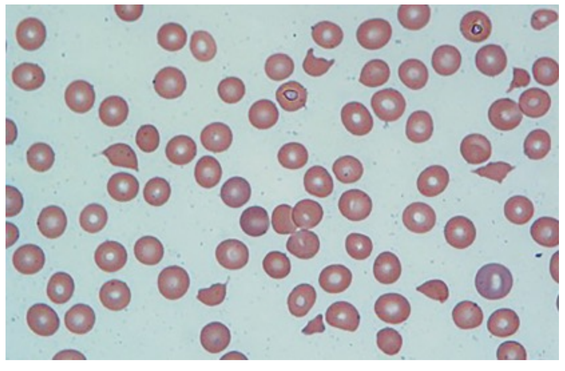

Figure 1: Schistocytes of MAHA

Case Presentation

A 32-year-old female with a chief complaint of bilateral lower extremity swelling and is found to have hypertensive emergency typified by acute kidney injury with a creatinine of 23.0 on admit. Hemoglobin on admit was 7.9 without a recent baseline value on record.

Initial workup into the cause of her normocytic anemia was undertaken with query into hemolysis as one of the possibilities. The morning following admit, the patient's hemoglobin was 5.6 with a low haptoglobin, high LDH, and schistocytes present on peripheral smear (see table 1). An initial diagnosis of MAHA was made. A critical point in the examination of this process had been arrived at: what was the cause of this MAHA?

Laboratory Values

Hemoglobin: 7.9 on admit, 5.6 on day 1

Blood smear: schistocytes present (see figure 1)

LDH: 532

Haptoglobin: 8

Platelets: range 89 – 107

Creatinine: 23.0 on admit

ADAMSTS-13: within normal limits

C3&C4: within normal limits

ANA: negative

Nephrotic range proteinuria

Table 1: Select Lab values over course of 7-day hospitalization

Investigation into the etiology of MAHA requires prompt investigation of concerning and rapidly progressive etiologies as these may result in significant increases in morbidity and mortality. A standard approach to separating the etiologies of MAHAs is into broad categories of inherited versus acquired.

The rapid onset of this anemia effectively rules out inherited causes. Acquired causes are separated into categories including infectious, toxic, autoimmune and mechanical.[6]

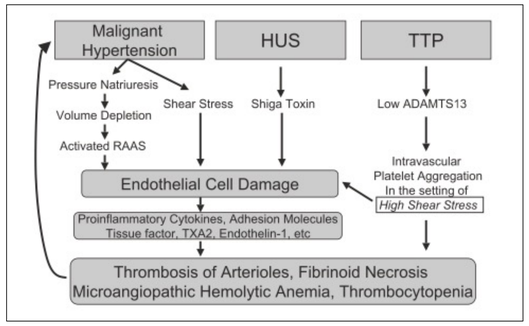

Figure 2: Pathogenesis of Malignant Hypertension, HUS and TTP in causing MAHA[4]

The confluence of profound hypertension and kidney injury provided further narrowing into a subset of autoimmune and mechanical causes including HUS, Malignant hypertension, and TTP. Figure 2 shows the pathogenesis of these particular etiologies while figure 3 suggests an approach based on platelet count in the setting of severe hypertension.

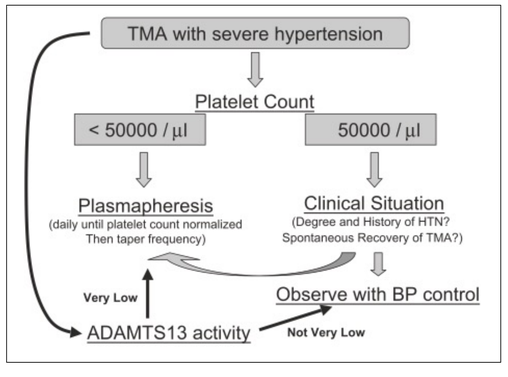

Figure 3: Proposed decision-making pathway in the approach and initial treatment of MAHA with severely elevated blood pressure and kidney injury[4]

Discussion

After ruling out causes through appropriate laboratory investigation (DIC panel, ADAMSTS-13, E coli O157-H7), a diagnosis of malignant hypertension-induced thrombotic microangiopathy was made. Further elucidation of the patient's overall presentation was made when kidney biopsy revealed AA amyloidosis as the cause of her proteinuria associated lower extremity swelling; the renal dysfunction associated with AA amyloidosis was a contributor to her progressive hypertension which then further hastened her progression to end-stage renal disease.

While the diagnosis of the usual offenders of combined kidney injury and hemolytic anemia (DIC, TTP, HUS) was absent, this case serves to illustrate the importance of investigation into the underlying cause of MAHA and its implications.

References

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Contact

Bryan.Takisaki@providence.org