AN INCIDENTAL FINDING OF TTP

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INTRODUCTION

Thrombotic thrombocytopenic purpura (TTP) is a rare thrombotic microangiopathy that classically presents with a pentad of microangiopathic hemolytic anemia, thrombocytopenic purpura, neurological dysfunction, fever and renal disease. This pentad is only present in about 40% of cases which makes diagnosis of this rare condition challenging especially in cases with pre-existing conditions. The onset is usually acute to sub-acute. It is often idiopathic but on rare occasions can be related to connective tissue diseases such as SLE. Making the early diagnosis of TTP is crucial as this condition has a 90% mortality rate if left untreated.

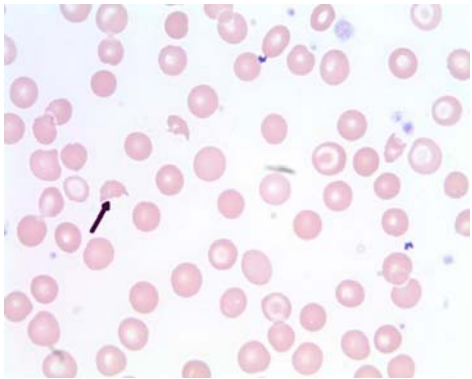
CASE PRESENTATION

A 36 y/o F with a past medical history of bilateral pulmonary embolism, systemic lupus erythematosus, and bipolar disorder presented to ED with an undetectable blood pressure in the outpatient setting which was quickly ruled out once she presented to the emergency department. Incidentally, her lab work was consistent with acute kidney injury (creatinine of 2.5mg/dL), anemia (HGB of 7.0mg/dL), and thrombocytopenia (15k/mcL). The patient had no active complaints at the time of admission. She did not have a fever and had no overt signs of bleeding on physical exam. Neurological exam was normal, although patient appeared to be in a manic state given her history of poorly controlled bipolar disorder. She appeared restless, had pressured speech and was pacing in her room.

Upon further investigation, she was found to have worsening of proteinuria w/ urine protein creatinine ratio of 14,639 mPR/gCR. Other labs, including peripheral smear, LDH, Haptoglobin, Coombs test, d-dimer, fibrinogen, direct and indirect bilirubin and ADAMTS 13 activity and inhibitor were collected. LDH on admission was 1,434 Units/L. She also had schistocytes on peripheral smear. She was initially started on treatment for immune thrombocytopenic purpura due to preexisting autoimmune disease, but that diagnosis was ruled out given negative Coombs test.

About 4 days later, the ADAMTS 13 activity came back at <5% and she was subsequently started on plasma exchange therapy. In retrospect, this therapy should have been started at first presentation and should have been higher on the differential.

She was discharged with hematology follow-up and continued with outpatient plasma exchange. Subsequently, her hemoglobin returned to her prior baseline ~9mg/dL. LDH also normalized. As of yet, she has not had a recurrence of her TTP.



Schistocytes on peripheral
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CONCLUSION

TTP is a rare diagnosis however it should remain on the differential diagnosis of a patient with anemia and thrombocytopenia until completely ruled out. At times, diagnosis can be clouded by underlying autoimmune conditions such as SLE. Plasma exchange has reduced mortality related to TTP from 90% to 10-20%. It is important to combine information from the history and physical exam, complete blood counts, LDH and peripheral smears to make the diagnosis of TTP. The ADAMTS13 should mainly be used as a confirmatory test. Treatment should not be delayed while awaiting the results of this test as the consequences of delaying treatment can be life-threatening.

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