Thrombotic thrombocytopenic purpura: a rare case presenting with splenic infarction

Abstract
Microangiopathic hemolytic anemia (MAHA), thrombocytopenia, fever, renal failure, and neurologic symptoms comprise the cardinal features of thrombotic thrombocytopenic purpura (TTP). Etiologies can include medications, infections, cancers, or transplantation. However, recognition of thrombotic thrombocytopenic purpura can be difficult because of the variety of presentations and lack of specific diagnostic criteria. We are presenting a case of acute TTP following a bout of splenic infarction. This rare case reminds us that splenic infarction can be an atypical presentation of TTP. Prompt recognition of TTP is important because the disease responds well to plasma-exchange treatment which improves patient’s prognosis, and is associated with a high mortality rate when untreated.

Keywords: thrombotic thrombocytopenic purpura, splenic infarction, microangiopathic hemolytic anemia (MAHA)

Introduction
Thrombotic Thrombocytopenic Purpura (TTP) is a hypercoagulable state, in which platelets aggregate and clot in the microvasculature. The majority of cases can be linked to ADAMST13 deficiencies, a protein responsible for cleaving VonWillibrand’s Factor (vWF). Hypofunctional ADAMST13 results in un-mitigated vWF activity, leading to excessive platelet aggregation. Increased clotting leads to decreased circulating platelets, hemolysis and anemia. Microvascular infarctions cause non-blanching purpura, renal damage, and cerebral ischemia resulting in a myriad of Central Nervous System manifestations from altered mental status to seizures. While some cases are idiopathic, 10% of TTP diagnoses are associated with sepsis or malignancy as well as enterocolitis, especially from E. coli. Originally, TTP was characterized by a classic pentad of microangiopathic anemia, thrombocytopenia, fever, neural manifestations, and renal damage. However, too many cases were missed so a more liberal diagnostic triad was created: microangiopathic anemia associated fragmented erythrocytes, and thrombocytopenia.

Case description
79 year old Hispanic male with past medical history of HTN and osteoarthritis presented to the emergency room complaining of left side abdominal pain for three days and intermittent confusion. Physical examination was unremarkable apart from fluctuating mental status and left lower quadrant tenderness on palpation of abdomen. Initial laboratory investigations revealed anemia, thrombocytopenia, elevated bilirubin but normal liver enzymes and lipase. The peripheral blood smear showed schistocytes. Imaging studies including abdominal Computed Tomography (CT) scan demonstrated a wedge-shaped hypo-enhancing lesions in the spleen typical for splenic infarcts as shown in Figure 1, and a brain CT scan was unremarkable.

Plasma exchange was initiated immediately along with systemic steroids. Platelets counts improved, the patient remains afebrile and hemodynamically stable but his altered mental status persisted. Further workup for his persistent change in mental status revealed multiple bilateral acute lacunar infarcts on Magnetic Resonance Imaging (MRI) of the Brain as shown in Figure 2, which is likely embolic in etiology. After six days, the patient became lethargic, febrile, tachycardia and the platelets count dropped again. Plasma exchange subsequently stopped when positive blood cultures grew a gram-negative bacteria (E. coli). The patient progressively improved following therapy with cefepime and gentamycin. Plasma exchange resumed and he maintained acceptable platelet count until being discharged from the hospital.

Figure 1
Computed Tomography scan of the Abdomen showing wedge-shaped hypo-enhancing lesion in the spleen.

Figure 2
Magnetic Resonance Imaging (MRI) of the brain showing multiple bilateral acute lacunar infarcts.
Twenty percent of patients will not respond to plasma exchange, making our first line treatment significantly flawed. On a side note, it is postulated that the majority of patients who do not respond to plasma exchange, developed TTP from atypical etiologies such as malignancy. As a second line treatment and for severe cases, immuno suppressants are used. The mainstays of this treatment are steroids and rituximab. Response to treatment with rituximab is usually seen within fourteen days. After platelets have normalized, treatment should be continued for at least 2 more days to insure efficacy. Given the intensity of the treatment, it is not surprising that 26% of patients develop major complications, including infections, venous clots, and hypotension. Two percent develop life-threatening conditions such as catheter-site hemorrhage, and catheter-related sepsis.

TTP is a rare but potentially fatal disorder, prompt recognition is important because the disease responds well to plasma-exchange treatment, but is associated with a high mortality rate when untreated. In the era before effective treatment with plasma exchange, 90 percent of patients with thrombotic thrombocytopenic purpura died from systemic microvascular thrombosis that caused cerebral and myocardial infarctions and renal failure. However, recognition of thrombotic thrombocytopenic purpura can be difficult because of the variety of presentations and lack of specific diagnostic criteria.

Conclusion

Our patient presented with abdominal pain secondary to a splenic infarct. Eighty-eight percent of splenic infarcts are caused by hematological problems such as blood cancers, hypercoagulable states, atrial fibrillation and vasculitides. However, very few have reported TTP as a precipitant. Indeed, this case is remarkable as a rare disease with an even more rare presentation. Given the severity, the patient was started immediately on steroids and plasma exchange. The brain MRI revealed one of the dreaded complications listed above: multiple, bilateral lacunar infarcts. Finally, our patient also developed a feared, predictable life threatening complication to the therapy–nosocomial E. Coli sepsis. Despite the myriad of set-backs, our patient improved and was discharged home.

This case stands apart as a rare presentation of a rare disease. Furthermore, it is a classic example of the importance of vigilance in treating a complex condition, with complications from both the disease, and the treatment. Early identification of TTP and anticipation of the adverse effects of treatment are vital in avoiding the significant morbidity and mortality of this potentially debilitating disease.

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Conflict of interest

The authors have no conflicts of interest to declare relevant to this article.

References


