

Incomplete Thrombotic Thrombocytopenic Purpura (TTP) with Stroke and Initial Negative Head CT

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ABSTRACT

A 52 year old African American female presented to the Emergency Department for the evaluation of left sided hand and facial weakness. She was noted to have a normal non-contrast head CT; however she did have thrombocytopenia on her complete blood count. Thus, initially the patient had microangiopathic hemolytic anemia (MAHA), thrombocytopenia and a neurological change consistent with thrombotic thrombocytopenia purpura (TTP). Subsequently, the patient developed the TTP pentad of MAHA, renal failure, thrombocytopenia, fever and neurological involvement. Thus the initial presentation was incomplete for the pentad of TTP. This case illustrates that the emergency physician must be on high alert for a patient who presents with neurological symptoms and thrombocytopenia despite not having the complete pentad for TTP.

KEYWORDS

Thrombotic Thrombocytopenia Purpura (TTP); Incomplete Thrombotic Thrombocytopenia Purpura (TTP); Emergency Department; Thrombotic Thrombocytopenia Purpura (TTP); Thrombotic Thrombocytopenia Purpura (TTP); Stroke.

INTRODUCTION

TTP is a rare condition which carries a mortality rate of > 90% if left untreated. Early treatment with plasma exchange can reduce the mortality to 10-20% if instituted within 4-8 hours of presentation [1]. Plasma exchange should be started if there is evidence of microangiopathic hemolytic anemia (MAHA) and thrombocytopenia with no other known cause.

CASE REPORT

A 52 year old African American female presented to the ED for evaluation of left sided hand and facial weakness which started approximately 30 minutes prior to her arrival in the ED. A stroke alert was immediately called. She was immediately taken down for a non-contrast head CT scan. The CT scan showed no acute pathology. On further evaluation, the patient report

ed that she had a history of multiple strokes in the past with a recent stroke approximately 1.5 months prior to this ED visit. Given this history of multiple strokes and a normal head CT with no evidence of prior stroke this seemed abnormal. There was no evidence of old infarcts that we could view. Neurology recommended that she be admitted to the hospital and a routine stroke workup should be undertaken. The initial platelet count was 74,000. Shortly thereafter, her platelet count was noted to be 44,000. The laboratory was then called and a smear was requested. The smear showed +2 schistocytes. After obtaining that information LFTs, LDH and a reticulocyte count were added to the requested labs. The BMP showed normal creatinine of 0.99. She had no fever. A call was placed to hematology to discuss the case. That opinion of the consul-

tant was that TTP was the likely diagnosis, given evidence of microangiopathic anemia as evidenced by schistocytes, along with the elevated LDH and reticulocyte count, in association with the neurologic complaints noted. Hematology recommended that the patient be admitted to the ICU and that plasmapheresis should be initiated. The patient was admitted to the ICU. She was also on Coumadin which she reported was for "irregular heart beat." Her INR was 2.1. Later the next afternoon she was noted to have an acute change in mental status. The neurologist then reevaluated her and recommended that she be transferred to a Neuro ICU. There she had a MRI which showed small infarcts in the left corona radiata and left parietal cortex. No acute neurosurgical intervention was needed. She then also developed a fever of 100.6 as well as an acute increase in her creatinine. Her maximal creatinine was 6.31.

Initially the patient only had a microangiopathic hemolytic anemia and a neurological change consistent with TTP. Later she developed the pentad of MAHA, renal failure, thrombocytopenia, fever and neurological involvement. Plasmapheresis was initiated early; approximately 14 hours after the patient was admitted to the hospital. The patient required approximately 30 days in the hospital with multiple rounds of plasmapheresis and hemodialysis to clear her pathology. She was then discharged to a rehabilitation facility, with minimal left sided residual deficit but off hemodialysis. Shortly thereafter, she presented to the hospital with a decreased platelet count and required repeat plasmapheresis. Lab testing was noted to have a decreased ADAMTS13 level. The patient had no history of recent diarrhea or of any new medications. It was felt that the ADAMTS13 level was a mutation that was unmasked as an adult.

DISCUSSION

The incidence of TTP is estimated to be between 2-15 cases per 100000 person years [1]. Many patients will initially only present with 2-3 elements of the TTP pentad of MAHA, renal failure, thrombocytopenia, fever and neurological involvement. The diagnosis does not require all five elements. This has been referred to in the literature as an incomplete form of TTP [2]. The pathogenesis of TTP has been hypothesized to be related to a vascular endothelial cell injury. This injury is thought to lead to the release of unusually large von Willebrand factor multimers. Platelet aggregation is induced by the action of these multimers on various platelet receptors [3]. Such multimers are usually cleaved by an enzyme known as ADAMTS 13. In most cases of familial or acquired TTP, the ADAMTS 13 enzyme is deficient, with low activity [4]. In some cases the ADAMTS 13 level may be undetectable [5].

This case the patient presented in an incomplete fashion, with MAHA, thrombocytopenia and neurological complaints. Further along in her hospital course, she progressed to the complete pentad where she became febrile and had acute renal failure requiring hemodialysis.

The diagnosis was supported by a blood smear, which the emergency physician requested from the laboratory, on which schistocytes were identified.

The relationship of TTP to stroke is complex. TTP may mimic acute ischemic stroke [6]. TTP may cause acute ischemic stroke [7]. A stroke may be the first clinical presentation of several hematological disorders, including TTP [8]. Thus, the relationship of TTP to stroke is complex. As noted by Burris et al, overall TTP is a rare diagnosis with frequent neurological involvement [9]. Stroke is one of possible neurological manifestations of TTP.

Of interest in the case also is that the patient's TTP resolved after approximately 30 days in the hospital but her symptoms returned shortly thereafter requiring further plasmapheresis. Such relapses have been described in the literature [2,3].

CONCLUSION

This case illustrates that the emergency physician must be on high alert for a patient who presents with neurological symptoms and thrombocytopenia despite not having the complete pentad for TTP. Many patients will initially only present with 2-3 elements of the TTP pentad of MAHA, renal failure, thrombocytopenia, fever and neurological involvement. The diagnosis does not require all five elements.

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