Thrombotic Thrombocytopenic Purpura or Disseminated Intravascular Coagulation? Diagnostic Dilemma in the ICU

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DIC and TTP are two causes of thrombocytopenia that require timely diagnosis and different treatments. Both conditions can be difficult to recognize as clinical presentations vary and current diagnostic criteria lack specificity. DIC is a complex thrombo-hemorrhagic condition that is always secondary to an underlying disorder, the most common causes being sepsis or trauma. It is primarily a clinical diagnosis that must be confirmed by laboratory data (see figure 2). There is, however, no single laboratory test that can establish or exclude the diagnosis. TTP is a rare condition characterized by systemic microvascular thrombosis, with an incidence of 4 to 11 cases per million people. Like DIC, TTP has no specific diagnostic test and it shares many of the clinical and laboratory features of DIC that can make the two diagnoses difficult to differentiate. Prompt recognition of TTP is warranted as it responds well to plasmapheresis, and without treatment it is associated with a high mortality rate. Although clinical presentation and laboratory data often lead to the correct diagnosis, equivocal results can often precede finding a clear etiology.

**Hospital Course**

This is the case of a 77 year old female with PMH of spinal degeneration ± multiple spinal surgeries, hypertension, and hypothyroidism, who presented to the surgical ICU with T9 ileum posterior fusion transverse osteotomy, L1-L2 transfemoral lumbar interbody fusion, and T11-T12 posterior laminectomy. Her operative course was prolonged due to an incidental durotomy. She required a phenylephrine infusion for the majority of the 11-hour and fluid resuscitation of 3 units of PRBCs, 7700 mL of crystalloid, and 780 mL of cell saved. Blood loss was estimated at 1500 mL and urine output was 1150 mL. Postoperatively she went to the ICU hemodynamically stable and intubated. By HD2, her renal function had worsened and she had developed thrombocytopenia (figure 1). A FENa done at the time was consistent with intrinsic renal damage. While awaiting WooCommerce and evaluated on HD2, her renal function continued to worsen and her platelets continued to drop. H&H and gaseous were ruled out, but a peripheral blood smear obtained on HD3 showed microangiopathic hemolytic anemia, turning the differential diagnosis to TTP or DIC. By then, the patient was also noted to have a nasogastric tube occluding an antecubital IV site. A DCC index done on HD4 was inconclusive, having revealed an elevated FDP and D-dimer with a low AT III but an elevated fibrinogen and marginally elevated PT and PTT. TTP was still being considered in the setting of renal failure which was likely acute tubular necrosis. On HD6 the patient developed uricemic encephalopathy and she required dialysis. After the patient’s platelets dropped to 21, plasma exchange was scheduled for HD7, but before initiating this treatment her platelets began to improve. The fact that her thrombocytopenia improved without intervention ruled out TTP as the etiology. She was transferred to the floor on HD8 with a diagnosis of DCC and AKI, likely due to an intra-operative event.

**Diagnostic Dilemma**

<table>
<thead>
<tr>
<th>TTP</th>
<th>DIC</th>
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<tbody>
<tr>
<td><strong>Diagnostic Criteria</strong></td>
<td><strong>Pathophysiology</strong></td>
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<tr>
<td>Unexplained microangiopathic hemolytic anemia and (2) thrombocytopenia (with or without neurologic involvement, renal failure, and fever)</td>
<td>Severe deficiency of von Willebrand factor causing platelet aggregation as a result of mutation in the ADAMTS13 gene leading to systemic microvascular thrombosis</td>
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<tr>
<td>Unexplained microangiopathic hemolytic anemia</td>
<td>Autoimmune hemolytic anemia,</td>
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<td></td>
<td>DIC, ITP, (2) thrombocytopenia, (3) prolongation of clotting times, (4) presence of fibrin degradation products, (5) low levels of coagulation inhibitors</td>
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</table>

**References**


**Abbreviations**

- HD = Hospital day
- TTP = Thrombotic thrombocytopenic purpura
- DIC = Disseminated intravascular coagulation
- AKI = Acute kidney injury