Learning objectives
- Understand the clinical presentation of TTP
- Be aware of reported cardiopulmonary complications associated with thrombotic microangiopathies and their mechanisms

Introduction:
- Thrombotic Thrombocytopenic Purpura (TTP) is characterized classically by neurologic changes, fever, renal failure, thrombocytopenia, and microangiopathic hemolytic anemia (MAHA)3,4.
- Only thrombocytopenia and MAHA are required for diagnosis in the correct clinical context.
- It is rarely accompanied by cardiopulmonary complications.

Case Information:
A 29-year-old woman with a history of chronic abdominal pain was brought to the Emergency Department. The patient had a history of one week of diffuse abdominal pain, diarrhea, progressing to vomiting and then to lethargy and altered mental status.

On presentation:
- Physical exam was notable only for dilated pupils and general confusion in response to questions
- At the time of admission, significant laboratory results are as reported in Table 1.
- A peripheral blood smear showed >10% schistocytes/hpf

Hospital Course:
- Treated with plasma exchange, steroids, hemodialysis, and intermittent blood transfusions for TTP
- On HD#7, she was found to be hypoxic and in acute respiratory distress and a CXR at the time (Image #2) was concerning for bilateral pulmonary edema
- Admitted to ICU, required NPPV
- BNP at the time was 4013.
- Transthoracic echocardiogram:
  - Ejection fraction of 40%
  - No left akiensis
  - PA systolic pressure of 48 mm Hg
  - She was treated with both continued hemodialysis and furosemide, afterload reduction with hydralazine
  - By HD #9, pulmonary edema had improved considerably, no longer had respiratory distress.

Discussion:
- The pathophysiology of TTP is thought to be due to the persistence of ultralarge vWF multimers which cause platelet aggregation and thrombi3,4.
- In autopsies, vWF-rich hyaline thrombi are found in arterioles and capillaries of various organs, including the heart, kidney, and brain5.
- Prior reports of cardiopulmonary complications:
  - Non-cardiogenic pulmonary edema: In a review of TTP cases from 1981-1998, 7 out of 56 cases of TTP were found to have ARDS, and in these cases, when plasma exchange was initiated, there was improvement of ARDS. The mechanism for ARDS in these cases was postulated to be microthrombi in pulmonary alveolar capillaries leading to hypoxemia and pulmonary edema1.
  - Takotsubo's cardiomyopathy: A case has been reported of a patient who developed acute cardiomyopathy with TTP, presumably from a stress response8.

In this case:
- Patient met 4/5 diagnostic criteria for TTP upon admission, lacking only fever.
- Respiratory distress differential included cardiogenic pulmonary edema and TRALI (given frequent transfusions). However, she developed LV dysfunction, and pulmonary edema resolved quickly with appropriate treatment of heart failure, suggesting that respiratory distress was more likely cardiogenic.

Clinical importance:
While respiratory dysfunction infrequently accompanies thrombotic microangiopathies, respiratory status should be monitored vigilantly as these patients are susceptible to both cardiogenic and non-cardiogenic pulmonary edema.

References:

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Image #1: on admission
Image #2: HD#7

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