MAYO CLINIC

19-Year-Old Woman With Multiorgan Failure and Purpura

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See end of article for correct answers to questions.

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A previously healthy 19-year-old female college student presented to her local emergency department for evaluation of fatigue, malaise, fever, and flulike symptoms including nausea and vomiting that had started the night before. She was managed with intravenous fluids and antiemetics and discharged home. However, she returned the following day with the same constellation of symptoms. She was febrile (temperature, 38.5°C), and laboratory test results were notable for mild hyponatremia (sodium, 132 mmol/L) and hypokalemia (potassium, 2.8 mmol/L). She was admitted for further observation. Her condition deteriorated rapidly, however, with evidence of hypotension, anuria, and increased oxygen requirements. Over the next hour, a rash developed on both lower extremities. Given this evidence of decompensation, the patient was immediately transferred to our medical intensive care unit for further evaluation and management. She was intubated and mechanically ventilated for acute hypoxemic respiratory failure and required vasopressors to maintain her blood pressure. Skin examination revealed a diffuse, maculopapular purpuric rash that involved her trunk and extremities. These lesions progressed to skin necrosis and gangrene over the course of a few hours. Results of a neurologic examination were notable for somnolence, but no focal deficits were evident.

1. On the basis of the clinical findings thus far, which *one* of the following is the *most likely* diagnosis?

a. Henoch-Schönlein purpura (HSP)
b. Thrombotic thrombocytopenic purpura
c. Allergic skin reaction
d. Purpura fulminans (PF)
e. Postinfectious thrombocytopenic purpura

Henoch-Schönlein purpura is certainly a consideration for this patient's rash; however, HSP rarely leads to skin necrosis and is not usually associated with the hemodynamic compromise seen in our patient. Thrombotic thrombocytopenic purpura can present with similar skin findings, but it never leads to skin necrosis. Although an allergic skin reaction is a concern, allergy-associated lesions tend to be more erythematous as opposed to purpuric and are typically pruritic. Purpura fulminans is associated with a diffuse rash that affects the whole body, and onset is 12 to 24 hours after initial flulike symptoms commence. It progresses rapidly to multiorgan failure, as seen in our patient, because of thrombotic occlusion of small and medium-sized blood vessels. Postinfectious thrombocytopenic purpura can have similar findings but rarely presents with shock. Our patient's findings are most consistent with severe acute infectious PF.

On admission, laboratory studies revealed the following: leukocytes, 29.8×10^9/L; platelet count, 52×10^9/L; sodium, 128 mmol/L; potassium, 5.0 mmol/L; bicarbonate, 12 mEq/L; creatinine, 2.7 mg/dL; international normalized ratio, 2.4; fibrinogen, 162 mg/dL; and elevated transaminase levels. The abnormal coagulation findings were consistent with disseminated intravascular coagulation (DIC). The lactate level was initially 4 mmol/L and peaked at 7.3 mmol/L. Arterial blood gas analysis revealed severe acidemia with a pH of 7.01, PCO_2 of 48 mm Hg, PO_2 of 95 mm Hg, and HCO_3 of 11 mmol/L. Chest radiography showed perihilar infiltrates with a consolidation in the right lower lobe. Bilateral pleural effusions and atelectasis in the mid left lung were also noted.

2. To further evaluate the patient's symptoms and to confirm the diagnosis, which *one* of the following would be the *most appropriate* next step?

a. Head computed tomography
b. Blood cultures and cerebrospinal fluid examination with rapid administration of broad-spectrum antibiotics
c. Laboratory tests for thrombocytopenia
d. Skin biopsy
e. Laboratory tests for vasculitis

The patient's deteriorating condition included altered level of consciousness and

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somnolence, which prompted concern about maintaining a patent airway. Although head computed tomography is certainly an appropriate diagnostic test for altered level of consciousness, her neurologic examination revealed no focal deficits and did not suggest a mass lesion or hemorrhage. Furthermore, her clinical presentation of fever and multiorgan failure are more suggestive of an infectious etiology. Given the concern for PF and the likely infectious etiology, the most appropriate next step in evaluation is blood cultures and cerebrospinal fluid examination, followed by immediate administration of broad-spectrum antibiotics. However, the administration of antibiotics should not be delayed if the tests cannot be performed immediately. Additionally, her laboratory evaluation had already revealed thrombocytopenia and other findings consistent with DIC. Skin biopsies can be useful in determining the etiology of rash. However, results are not immediately available, and essential treatment should not be delayed in this critically ill patient. Because the leading diagnosis is PF, skin biopsy would not add useful information. Laboratory tests for vasculitis could be helpful if the suspected diagnosis was HSP but would not aid in the diagnosis and management of PF. Our patient is a young woman attending college whose overall presentation is concerning for PF secondary to an infection based on her skin findings, multiorgan failure, and shock.

3. Which *one* of the following is the *most likely* cause of this patient's PF?

a. Protein C or S deficiency
b. *Staphylococcus aureus*
c. *Neisseria meningitidis*
d. *Haemophilus influenzae*
e. *Streptococcus pneumoniae*

Several infectious and noninfectious etiologies have been implicated in PF. One noninfectious etiology is protein C or S deficiency, which can result in PF in patients receiving warfarin therapy. Our patient was not taking anticoagulants. Rarely, *S aureus* can cause PF. *N meningitidis* is the most common cause of PF. Given the patient's age and status as a college student, this is the most likely etiology. *H influenzae* and *S pneumoniae* can cause PF but are less likely in this patient.

Blood and cerebrospinal fluid analysis confirmed the presence of *N meningitidis* in our patient with septic shock and multiorgan failure from PF.

4. Which *one* of the following is the *best* immediate treatment strategy for this patient?

a. Early goal-directed therapy (EGDT) with aggressive fluid resuscitation and broad-spectrum antibiotics
b. Antibiotics alone
c. High-dose corticosteroids alone
d. Transfusion of blood products
e. Immunoglobin therapy along with high-dose corticosteroids

This patient is acutely ill with septic shock. Early goal-directed therapy with aggressive fluid resuscitation along with rapid administration of antibiotics is required to treat her septic shock and underlying infection. Early goal-directed therapy improves mortality in patients with severe sepsis and septic shock. Antibiotics alone would not be adequate. Although corticosteroids can be used in patients with severe refractory sepsis/septic shock to help address the hemodynamic instability when vasopressors are used, this approach would not address the underlying infection. Corticosteroids should not replace intravenous fluids, antibiotics, and vasopressors. Although corticosteroids are used in patients with pneumococcal meningitis, they add no benefit in patients with meningococcal meningitis. Because our patient has no evidence of vasculitis, high-dose corticosteroids are not required. Transfusion of blood products is certainly indicated considering the degree of DIC in this patient, but it does not suffice as stand-alone management. Immunoglobulin therapy can be used in patients with thrombotic thrombocytopenic purpura or posttransfusion purpura, but this was not the diagnosis in our patient.

Although EGDT was instituted in our patient, her course was further complicated by adrenal insufficiency and acute renal failure requiring dialysis. The patient's initial creatine kinase level was elevated at 1032 U/L on admission and peaked at more than 20,000 U/L on hospital day 4, confirming the presence of compartment syndrome affecting her upper extremities. Bilateral forearm fasciotomies performed on hospital days 2 and 3 revealed