Purpura Fulminans: An Ominous Sign of Disseminated Intravascular Coagulation and Sepsis

Rishi Sharma,* Imran Ahmed, DO,* Andleeb A. Usmani, DO**

*Medical Student, 3rd year, Kansas City University College of Osteopathic Medicine, Kansas City, MO
**Dermatologist, Wellington Medical Center, Wellington, FL

Disclosures: None
Correspondence: Rishi Sharma; rishigautam500@gmail.com

Abstract

Purpura fulminans is a rare skin manifestation of disseminated intravascular coagulation (DIC) and may be secondary to sepsis. While the skin manifestations may not be the most emergent cause of death in septic patients – cause of death is typically shock due to decreased vascular resistance caused by endovascular toxins and bleeding from DIC – it is important to recognize this physical exam finding when parameters such as temperature, heart rate, and blood pressure are not readily available. We present a case of an assisted-living patient with purpura fulminans to illustrate the importance of recognizing purpuric skin lesions that could be consistent with PF.

Case Report

An 86-year-old female was brought by ambulance from an assisted living facility with complaints of altered mental status and weakness. The nursing staff at the facility had promptly called an ambulance due to her symptoms. En route to the emergency room, she was noted to have a blood pressure of 90/62 and a rectal temperature of 94 degrees Fahrenheit. Upon arrival to the ER, a sepsis alert was called.

The nursing facility was called for further history. They stated that the patient was having unusual skin findings for about three weeks, including a red, dry skin rash that started remotely to the chest and spread outward to the extremities over the next two weeks. More recently, she had developed purpuric lesions that were scattered on her hands and legs. For the rash, a dermatology consultation was requested, but it was not completed before her presentation to our hospital. A steroid cream was used by staff while awaiting dermatology consultation. Due to her unstable vital signs and the risk of possible sepsis, the patient was admitted to the critical care unit.

The patient’s white blood cell count was (21.8 x 10^9)/mm^3 with 78% segmented neutrophils, hemoglobin 13.5, hematocrit 42.3, and platelet count 170. Chemistry showed: sodium 141 mEq/L, potassium 4.9 mEq/L, chloride 108 mEq/L, bicarb 24 mEq/L, BUN 46 mEq/L, creatinine 2.08 mg/dL, glucose 100, calcium 8.3 mg/dL, total bilirubin 1 mg/dL, AST 65 U/L, and ALT 40 U/L. Total alkaline phosphatase was 177 U/L, albumin was 1.7 g/dL, and total protein was 6.2 g/dL. A coagulation panel showed: INR 1.7, PT 13.5 seconds, and PTT 35 seconds. A urine panel showed: cloudy urine with protein, positive blood, positive leukocyte esterase, 10 to 12 red blood cells, and 25 to 50 white blood cells, with a urine culture growing E. Coli. D-Dimer was 2585.

The patient’s vital signs were the indication of an emergent underlying cause. Hypothermia may be present in sepsis, especially if the temperature is less than 95°F. In addition, an elevated heart rate and low blood pressure are signs of shock. Altered mental status is one of the most important and easily overlooked signs of infection or other disruption to the patient’s homeostasis. The presence of these findings around the same time as a new eruption of rash should raise concerns and calls for prompt transfer to an emergency department.

Gram-negative sepsis is the most common cause of disseminated intravascular coagulation. The coagulation is caused by an inciting factor that damages the vasculature, such as lipopolysaccharides on the surface of the bacterial lipid membrane. This damage results in quick consumption of coagulation factors, resulting in an increased risk for bleeding. The perpetuation of coagulation by the offending agent results in increased production of thrombi consisting of fibrin and platelets, which may occur in both large vessels and microvasculature. Examples of large-vessel involvement include pulmonary embolism and thrombotic cerebral stroke, two causes of death in this setting. Microvascular involvement was demonstrated in our case, with necrotic skin...
lesions resulting in a nonpalpable, non-blanching, widespread purpura. This necrosis may result in sloughing of the skin with subsequent weeping of sanguineous fluid due to the increased susceptibility to bleeding. In addition, our patient had new and old areas of small petechiae, further demonstrating the clinical picture of DIC. This points to an infectious etiology for the patient’s purpuric lesions. Infectious purpura fulminans is more commonly encountered in the adult population.  

There are several subtypes of purpura fulminans other than infectious. Neonatal purpura fulminans is a rare, life-threatening disease caused by either a congenital or an acquired deficiency of protein C or protein S. The activated form of protein C, an antithrombotic protein derived from the liver, inactivates factors V and VIII, preventing excessive thrombus formation in the coagulation cascade. In the event of an acquired or congenital protein C deficiency, factors V and VIII are free to perpetuate thrombus formation, leading to micro- and macrovascular thromboses. Clinically, a neonate with congenital protein C deficiency may develop multiple well-defined, dark-purple to blackish patches involving random areas of the skin and extremities as soon as a few hours after birth. An acquired protein C deficiency may occur as a manifestation of sepsis, infection from gram-negative organisms and Staphylococcus species. Cases in which the etiology of the purpuric lesions is not clearly demonstrated may be termed “idiopathic purpura fulminans.” While not many cases of idiopathic purpura fulminans have been described in the literature, it reportedly presents with well-demarcated, purple to black patches anywhere on the skin in patients demonstrating relatively normal lab values, including normal prothrombin time, activated prothrombin time, protein C, protein S, and antithrombin III levels. Usually, idiopathic purpura fulminans affects the breasts or lower limbs, and it may arise up to 10 days after an antecedent infection, such as meningococcemia, Haemophilus, Staphylococcus aureus, or varicella.

**Conclusion**

The skin lesions found in purpura fulminans may be the result of microvascular thrombosis resulting in skin necrosis. It is a rare skin manifestation secondary to multiple causes, including disseminated intravascular coagulation, sepsis, warfarin skin necrosis in a setting of acquired protein C deficiency, heparin-induced thrombocytopenia, and thrombotic thrombocytopenic purpura. A new finding of dark, purpuric lesions in a patient demonstrating no identifiable trauma or other immediate cause calls for prompt evaluation by a health professional. Quickly attainable vital signs such as blood pressure, core temperature, heart rate, and an assessment of mental-status changes compared to baseline are helpful in identifying not only the severity of the skin findings but also the cause. Lab work demonstrating decreases in platelets, coagulation factors and hemoglobin, along with an increased D-Dimer, should prompt one to think DIC. A skin biopsy is helpful in ruling out vasculitides if the cause of the purpura is still uncertain. Appropriate management, including IV antibiotics, fluids, and adrenergics to treat a possible underlying sepsis, should be promptly initiated.

**References**