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# Purpura Fulminans: A Cutaneous Marker of Disseminated Intravascular Coagulation

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A previously healthy 14-year-old girl presented to the emergency department with high fever, cough, shortness of breath and right lobar pneumonia on chest radiograph. She had extensive purpura with hemorrhagic bullae on her left leg. The patient was very ill-appearing with hypotension, tachycardia, tachypnea, and oliguria. There was no other bleeding. Hemogram showed leukocytosis (13000/cmm) with 35% bands, platelets 76,000/ml and sedimentation rate of 98 mm. The prothrombin time and partial thromboplastin time were prolonged and the fibrin degradation products were grossly elevated. Blood culture grew group-A streptococci. A diagnosis of purpura fulminans from septic shock was made. She was resuscitated and given parenteral antibiotics and platelets. The patient recovered within two weeks and later had skin grafting.

Purpura fulminans is an infrequent, often fatal, acute cutaneous reaction resulting from infective or non-infective conditions.<sup>1</sup> When it arises during sepsis, in-hospital mortality is 42%.<sup>2</sup> Antecedent infections are most commonly group-A streptococcus, staphylococcus, pneumococcus, vibrio, and meningococcus, and less commonly varicella.<sup>3</sup> Neonates with protein C and protein S deficiencies are at higher risk for purpura fulminans. Patients with systemic lupus erythematosus may have antiphospholipid antibody syndrome.<sup>1</sup> The disease may occur without preceding illness.<sup>1</sup>

Purpura fulminans from sepsis requires surgical debridement, skin grafting and even amputation.<sup>4</sup> Normal saline resuscitation restores volume and promotes urine output >0.5ml/kg/hour.<sup>5</sup> Although there is no proven benefit, treatment of severe disseminated intravascular coagulation with purpura fulminans with heparin may be warranted.<sup>5,6</sup> Most clinicians prefer to provide platelet replacement if platelet counts drop below 20,000/mL.<sup>5,6</sup> Administration of protein C concentrate early in the course of the disease may reduce both morbidity and mortality.<sup>3</sup> There is some evidence that recombinant tissue plasminogen activator infusion may result in improved organ perfusion and cardiac performance.<sup>7</sup>



**Figure.** Extensive areas of purpura, ecchymosis and skin necrosis with hemorrhagic blebs and disrupted bullae, involving the left lower extremity.

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