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4-29-2020

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Recommended Citation

Grant, Leah and Plotinsky, Rachel, "Purpura fulminans due to MSSA Toxic Shock Syndrome" (2020).

Providence St. Vincent Internal Medicine. 3.

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Purpura fulminans due to MSSA Toxic Shock Syndrome



Leah Grant MD and Rachel Plotinsky MD

Introduction

Purpura fulminans is a rare and serious complication of an acute infectious process, characterized by large purpuric skin lesions, fever, hypotension, and DIC. The most common infectious cause of purpura fulminans is meningococcal disease, though few case reports in the literature describe *Staph aureus* as a causative organism.

Case Report

- A 53 year-old woman with a history of heart block s/p pacemaker placement in 2002 presented to the ED with nausea, vomiting, fevers, chills, diffuse muscle pain and a syncopal episode.
- She was found to have multi-organ dysfunction on labs, including AKI and thrombocytopenia. On hospital day 2, she developed acrocyanosis. She became febrile & hypotensive requiring transfer to the ICU for vasopressor support. Blood cultures were obtained. She was started on broad-spectrum antibiotic therapy.
- Because of the patient's thrombocytopenia and overall critical illness, the differential diagnosis included TTP, DIC, HUS, drug-induced hemolytic anemia, or a rare disorder called catastrophic antiphospholipid antibody syndrome (CAPS).
- She was treated empirically for CAPS with plasma exchange, heparin, and steroids.
- Blood cultures were positive for methicillin-sensitive *Staph aureus*, and antibodies for CAPS were negative.
- Antibiotic therapy was narrowed to Cefazolin, and her infected pacemaker was extracted.
- She required bilateral below-the-knee amputations as well as multiple finger amputations due to necrosis.
- Most likely diagnosis is purpura fulminans from MSSA toxic shock syndrome.



Discussion

- In acutely ill patients with skin findings described in this case as well as multi-organ dysfunction, there are several life-threatening diagnoses which must be recognized and treated promptly.
- Given the 50% risk of mortality even with prompt initiation of therapy for CAPS, we did not delay in starting this patient on plasma exchange.
- In purpura fulminans, the clotting cascade is disrupted by bacterial endotoxins and inflammatory cytokines, leading to a procoagulant and anticoagulant state, which in turn leads to intravascular thrombosis and hemorrhagic infarction of the skin.
- A report of 5 cases of purpura fulminans caused by MSSA TSS was published in the journal *Clinical Infectious Diseases* in 2005, and the isolated strains of MSSA were noted to produce higher-than-expected levels of endotoxins normally associated with TSS.
- Treatment of purpura fulminans from MSSA TSS is antibiotic therapy.
- It remains a rare and serious complication of acute infection which providers should keep on their differential of life-threatening illnesses associated with thrombocytopenia and purpuric skin lesions.

