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CASE REPORT
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Post-Cesarean Section Pyoderma Gangrenosum Presenting with Vasopressor-dependent Shock: Long-term Follow-up after Delayed Primary Closure

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Summary: A 28-year-old woman with poor wound healing and surgical site pain presented 5 days post-cesarean section (post-CS) with vasopressor-dependent shock and was eventually diagnosed with postoperative pyoderma gangrenosum (PG). A worsening clinical picture consistent with presumed necrotizing infection necessitated surgical debridement. The patient was ultimately taken to the operating room 4 times with transient improvement after the operations when she received perioperative corticosteroids. We were unable to identify an infectious source and cultures revealed no microorganisms. Dermatopathology revealed neutrophilic infiltrate and focal necrosis without microorganisms. The biopsy site began to concurrently exhibit pathergic changes, leading to a diagnosis of PG. Twelve weeks later, she underwent DPC of her abdominal wound while maintained on an immunosuppressive regimen of cyclosporine and prednisone. Incisional negative pressure wound therapy with a small window was used in the immediate postoperative period to allow for direct visualization of the closed incision. She healed without issue and her immunosuppressive regimen was ultimately discontinued. Postoperative PG is an uncommon diagnosis with high risk of morbidity. It is often mistaken for necrotizing infection. We report a unique case of post-CS PG presenting as vasopressor-dependent shock that was successfully closed with incisional negative pressure wound therapy with a small window. *(Plast Reconstr Surg Glob Open 2021;9:e3427; doi: 10.1097/GOX.0000000000003427; Published online 16 February 2021.)*

INTRODUCTION

Pyoderma gangrenosum (PG) is an ulcerative neutrophilic dermatosis with an annual incidence of 3–10 patients per million.[1,2] Acute onset PG often presents with a toxic picture, fever, and elevated inflammatory markers.[2] PG is commonly misdiagnosed as infection, but the lesions are culture negative.[3–5] Pathergy, the development of PG secondary to trauma, occurs in 20%–30% of cases and is considered pathognomonic.[3,4] Misdiagnosis of PG as infection often leads to surgical debridement, which inevitably exacerbates symptoms of PG and further delays definitive diagnosis and treatment.[6]

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PG has been associated with pregnancy, typically occurring in the second or third trimester or postpartum.[7] There are several reports of post-cesarean section (post-CS) PG, but it remains a rare phenomenon with ongoing controversy regarding surgical wound closure techniques.[7,8] We report a unique case of post-CS PG presenting as vasopressor-dependent shock and successful surgical intervention with delayed primary closure (DPC) while the patient was on immunosuppression.

CASE

A 28-year-old woman presented to an outside hospital with a surgical site infection 5 days post-CS. She was found to have a leukocytosis of 34,000 cells/mm^3. She continued to decline after initiation of broad-spectrum antibiotics and surgical debridement, necessitating transfer to our institution.

On presentation, she was febrile, tachycardic, and hypotensive, requiring vasopressor support. She had exquisite surgical site tenderness, a new oxygen requirement,

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increased leukocytosis (48,000 cells/mm^3), and lactic acidemia (2.4 mmol/L). We proceeded with urgent surgical debridement for presumed necrotizing infection (Fig. 1). Frankly necrotic skin and fascial edges were sharply debrided. Fascia was closed over a negative pressure wound therapy (NPWT) device; the skin was also partially closed. Dexamethasone (4 mg) was administered intraoperatively. Tissue cultures were negative. Tender red nodules, reportedly present since delivery, were noted on the extensor surfaces of her upper extremities and the inside of her left thigh.

She subjectively improved the following day, but her leukocytosis persisted (46,000 cells/mm^3). Dermatology evaluated the extremity lesions which were initially attributed to erythema nodosum in response to an infectious process. The patient deteriorated again on day of admission (DOA) 3 and her antibiotics were broadened. Pain limited examination, thus we returned to the OR. Improvement of the wound was noted so the fascia was closed and NPWT replaced under the partially closed subcutaneous tissue. An incisional biopsy of the lesion on her thigh was obtained. Perioperative steroids were not administered.

The patient continued to decline on DOA4. Operative exploration again failed to identify an infectious source. Dexamethasone (4 mg) was again administered intraoperatively followed by transient clinical improvement and subsequent deterioration on DOA7. Examination revealed biopsy site dehiscence and purulence. Rheumatology was consulted given this new finding.

On DOA8, we returned to the OR to change the NPWT dressing. Wound edge necrosis with a violaceous border surrounded by erythema and newly formed pustules were observed which raised suspicion for PG. The diagnosis was confirmed by pathology revealing neutrophil-rich infiltrate with organizing thrombi, focal necrosis, and abscess formation without microorganisms (**see figure, Supplemental Digital Content 1**, which displays the histopathologic specimen obtained from biopsy of lesion on the right thigh shown at 2.5× (1a) and 10× (1b) magnification showing neutrophil-rich infiltrate with organizing thrombi, focal necrosis, and abscess formation without microorganisms, **http://links.lww.com/PRSGO/B586**). Following intraoperative dexamethasone, intravenous methylprednisolone was initiated postoperatively. The patient rapidly improved and her leukocytosis resolved. She was transitioned to oral prednisone and nonadherent dressing changes upon discharge. While outpatient prednisone was tapered, cyclosporine was started, and wound care was transitioned to NPWT. Her wound continued to decrease in size and granulate without signs of PG (Fig. 2).

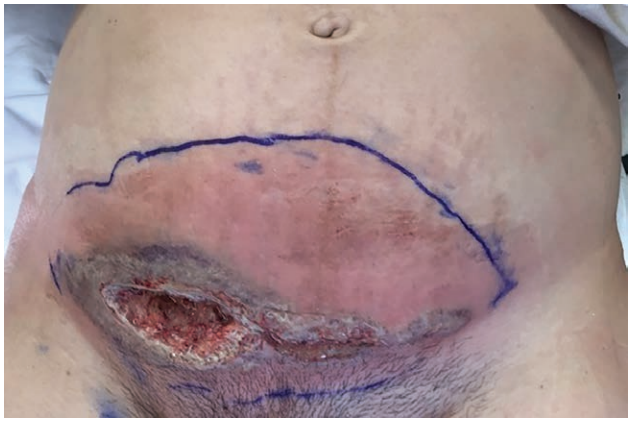

Fig. 1. Abdominal wound on presentation to our institution.

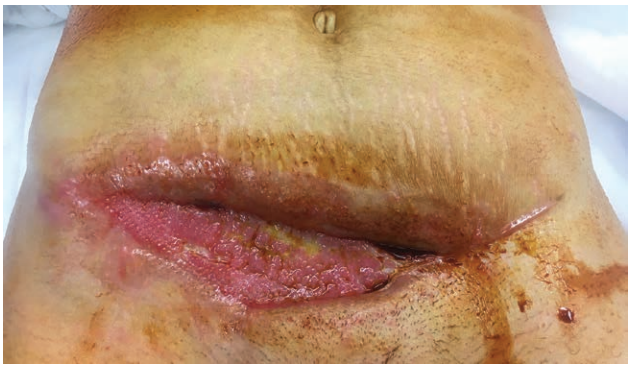

Fig. 2. Progression of wound during and after continued debridement. Abdominal wound (17 × 3.7 × 1 cm) demonstrating good granulation tissue without signs of PG after NPWT before DPC.

After multidisciplinary consultation, the patient was deemed ready for definitive closure 12 weeks post-CS as this time allowed for thorough workup and stabilization of the patient's PG. At the time of DPC, the patient was on cyclosporine and prednisone. The skin and subcutaneous tissue flaps were elevated to achieve tension-free primary closure with interrupted vertical mattress polydioxanone sutures and minimal instrumentation to reduce pathergy risk. Incisional NPWT with a 2 × 2 cm window in the dressing was used to further alleviate tension while allowing for direct visualization of the closure (Fig. 3A).

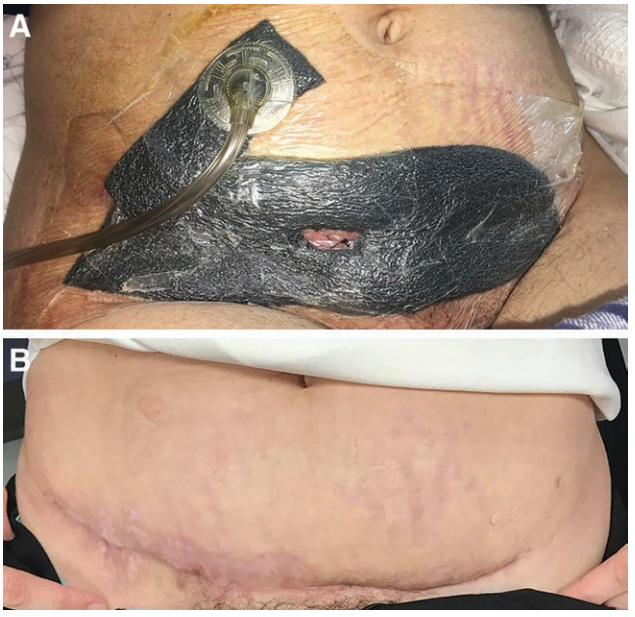

Fig. 3. Delayed primary closure of wound. NPWT with window to monitor closed incision (A). Complete healing of wound observed without recurrence of PG (B).

Daily laboratories were monitored for signs of a systemic response to new tissue trauma from wound closure. NPWT was removed on postoperative day 5. The wound remained well-approximated without pathergic changes. Immunosuppression was discontinued 6 weeks later without symptom recurrence (Fig. 3B).

The patient delivered her second child vaginally 17 months after DPC. She was admitted on postpartum day 8 with fever, leukocytosis, elevated inflammatory markers, and pelvic examination concerning for PG recurrence. Further workup and stable pelvic examination lowered the suspicion for PG and she was ultimately discharged on hospital day 2 followed by symptom resolution without antibiotics or steroid therapy. She remains off immunosuppression and has not experienced any symptoms concerning for PG at 18 months follow-up.

DISCUSSION

There are rare reports of PG presenting as shock one of which presented as vasopressor-dependent shock.[9] PG-associated shock is aseptic. It is believed that hypotension can develop as a result of an overpowering cytokine response triggered by severe PG.[9] A 2012 case report described a patient presenting with shock and tissue necrosis post-CS.[10] After diagnosing sweet syndrome, a neutrophilic dermatosis similar to PG, she was successfully managed with immunosuppressive therapy and split-thickness skin grafting.[10]

In our case, transient postoperative clinical improvement later attributed to perioperative steroid administration confounded our assessment of the utility of surgical debridement. Biopsy site pathergy was critical to diagnosing PG. Given the risk of further pathergy in PG, surgical intervention remains controversial and healing by secondary intention is typically favored. Split-thickness skin grafting and NPWT are also common approaches but may result in poor cosmesis.[7,11] If surgical intervention is pursued, sufficient immunosuppression and minimizing skin trauma are key to avoid further pathergy.[2,3,11]

DPC in our patient allowed for timely healing and improved cosmesis compared to healing by secondary intention. Our novel NPWT modification, a window for direct visualization of the incision, allowed for close monitoring of the incision and adjustments of immunosuppressive medications as necessary. Without this modification, the benefits of NPWT are far outweighed by the inability to directly visualize the incision.

Post-CS PG is uncommon and has a high risk of morbidity. Absence of underlying systemic disease and the severe, atypical presentation as vasopressor-dependent shock complicated our diagnosis. This case highlights the importance of pathergy to distinguish PG from necrotizing infection and avoid inappropriate surgical interventions. Optimization of immunosuppressive therapy and close monitoring of the incision with direct visualization are key to successful DPC.

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