BRIEF ARTICLES

Frequently debrided and misdiagnosed: Post-surgical pyoderma gangrenosum

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ABSTRACT

Post-surgical pyoderma gangrenosum is rare. On average it develops 10 days after a surgical procedure. This timeframe is similar to post-operative wound infections, including post-operative necrotizing fasciitis. Consequently, post-surgical pyoderma gangrenosum is frequently misdiagnosed as an infection, leading to detrimental surgical debridement, unnecessary antibiotic use, and delay of proper treatment. Herein, we describe a case of pyoderma gangrenosum of the right inguinal crease following percutaneous coronary catheterization and provide a literature review.

INTRODUCTION

In many surgical specialties, minimally invasive procedures have become the mainstay of therapeutic interventions. While this has decreased the rates of many post-operative adverse events, pyoderma gangrenosum in the post-surgical setting continues to occur and remains well documented (1, 2). Post-surgical pyoderma gangrenosum (PSPG) is characterized by an expanding surgical wound, painful central ulceration with an undermined, gray border and necrotic base. PG is estimated to occur in 3 to 10 cases per million population world-wide each year, and individuals aged 25 to 54 years are most frequently affected. (3). A female predilection has been reported for PSPG (2). This condition is frequently misdiagnosed as post-operative infection.

CASE REPORT

We report a case of an 80-year-old Caucasian female with multiple comorbidities, including a history of low grade, transfusion-dependent myelodysplastic syndrome (MDS) without additional treatment, hypertension, and chronic systolic congestive heart failure with an ejection fraction of 42%, who was admitted to the hospital for a myocardial infarction. She underwent cardiac catheterization through her right femoral artery, and within the week following the procedure, she developed a red papule at the procedure site which then ulcerated. She was treated for a presumed post-operative wound infection with multiple courses of IV vancomycin, piperacillin/tazobactam,

Figure 1: Right inguinal fold with deep, necrotic ulceration.

clindamycin, surgical debridement, and wound vac placement. Wound cultures were negative and the patient was afebrile throughout her hospital course.

The ulceration continued to enlarge. After four weeks, a second debridement was performed, and a biopsy was taken during debridement. Dermatology was consulted. Examination of the right inguinal fold revealed a large, deep, necrotic ulceration with a "vegetative" irregular, erythematous border (Figure 1). The ulcer and surrounding tissue were extremely painful. Histopathology revealed extensive ulceration in association with a central zone of necrotizing suppurative inflammation with sheets of neutrophils and inflammatory debris. A peripheral vascular reaction defined by perivascular and intramural lymphocytic infiltrates with a peripheral neutrophilic component, although without fibrinoid necrosis, was also noted (Figure 2). Special stains (Gram, PAS, AFB and Fite) were negative for gram +/- bacteria, fungi, and acid-fast mycobacterial organisms.

A diagnosis of post-surgical pyoderma gangrenosum was made. Intravenous 1 gram methylprednisolone was given for 3 days, followed by transition to 60 mg prednisone daily. In conjunction with cardiology, the decision was made to initiate infliximab 5mg/kg due to its fast onset of action and weight-based dosing. Cardiology deemed the patient's heart failure to be stable, and the rapid expansion of the ulceration was resulting in severe pain and suffering on behalf of the patient. The patient was transferred to a rehabilitation facility and died within three weeks. The cause of death was not determined.

DISCUSSION

Patients with PSPG are commonly admitted to a surgical subspecialty service. These services are accustomed to diagnosing and treating post-operative wound infections and other wound complications. PSPG initially presents as erythema encompassing an incision or suture lines, which develops into

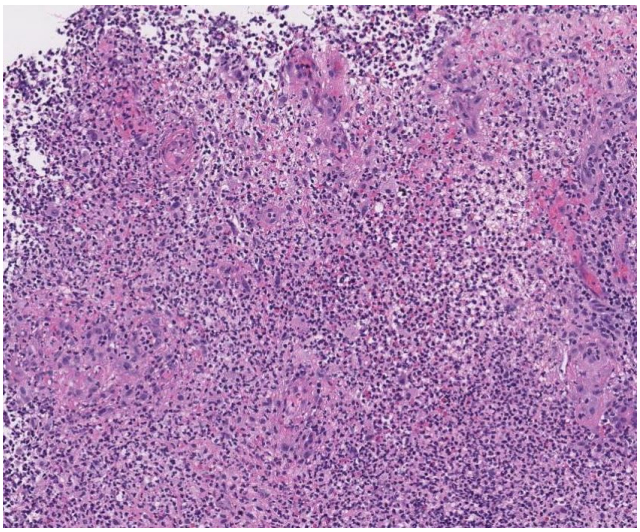

Figure 2: Histology showing perivascular and intramural lymphocytic infiltrates with a peripheral neutrophilic component, although without fibrinoid necrosis.

punctate ulcerations that later coalesce into a large painful ulcer with a rolled or undermined border. By contrast, necrotizing fasciitis tends to present as a patchy skin discoloration, usually dusky red or violaceous, with poorly-defined expanding erythema that develops into a tense edema with bullae, vesicles, or necrosis (4). Patients with necrotizing fasciitis may rapidly decline, becoming febrile, systemically unstable and septic. Both necrotizing fasciitis and PSPG are excruciatingly painful, which is out of proportion to their symptoms. When PSPG is mistaken for a wound infection, debridement, skin grafting, and local flap coverage may be undertaken, which induces a pathergic response that exacerbates the ulceration and causes expansion.

In a retrospective study of 18 patients with PSPG seen at the Mayo Clinic over a 20-year period, debridement was performed in 11 (61%) of the patients (2). PSPG should be suspected in the post-operative setting whenever the clinical findings of PSPG are present, painful ulceration occurs, and when ulceration is progressing in spite of antibiotic therapy and surgical debridement. A wound culture is highly unreliable because a negative culture does not rule out an infection while a positive culture may reflect low levels of bacterial colonization in the context of PSPG (5).

In our case, the patient's history of myelodysplasia served as another helpful diagnostic clue. Myelodysplastic syndrome (MDS) consists of heterogeneous clonal hematopoietic stem cell malignancies that are driven by immune dysregulation, and which confer increased risk of progression to acute myelogenous leukemia (6). While pyoderma gangrenosum (PG) is idiopathic, over 50% of patients with PG have a comorbid systemic disease, including MDS, inflammatory bowel disease, and rheumatoid arthritis (2,4,5). PSPG is associated to a lesser degree with systemic disease than other types of PG, however hematologic dyscrasias have been reported to be the most common condition in this subtype (2,4).

The distinction of PG from other ulcerative processes with dermal neutrophilia based on histology alone is challenging and, at times, impossible. Therefore, a detailed clinical history and solid knowledge of the underlying systemic disease or the associated processes is essential to reach the correct diagnosis. Histologically, PG may closely mimic Sweet's syndrome, which is a common manifestation of underlying hematopoietic disorders, however, clinical features make the distinction possible. As the lesions of PG are frequently follicular-based, other causes of necrotizing pustular follicular reactions with associated vasculopathy should be considered in the differential diagnoses. These include rheumatoid vasculitis, mixed cryoglobulinemia or Behcet's disease. However, these conditions are often associated with neutrophil-predominant necrotizing vasculitis in contrast to PG, which often shows a mononuclear-predominant non-necrotizing vascular reaction (7).

There is a dearth of evidence-based literature regarding effective therapies for PG. However, the use of immunosuppressive therapy with systemic corticosteroids and cyclosporine, either individually or in combination, is well established as first line therapy in the literature (2). Effective treatment of PG with topical corticosteroids and tacrolimus has also been reported, but these are usually restricted to limited and localized cases of PG (8,9). Dapsone has demonstrated efficacy in the treatment of PG in children, with a success rate of approximately 80% (10). Effective suppression of pathergy using anti-tumor necrosis factor agents such as infliximab,

etanercept, and adalimumab have also been reported (11,12). Alternative treatment options include hyperbaric oxygen, methotrexate, cyclophosphamide, mycophenolate mofetil, sulfasalazine, and azathioprine (1).

PG has a good prognosis when diagnosed and treated immediately; however, if appropriate therapy is delayed, the prognosis worsens and may lead to sepsis and death (5). While PG is well documented in the dermatology literature, it is less frequently reported in the surgical or cardiology literature. This underscores the importance of increasing awareness of PG as a differential diagnosis for wound infections among non-dermatology specialists and surgeons. As this condition can be easily misdiagnosed and incorrectly treated, early consultation with dermatology is highly recommended

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References:

1. Almukhtar R, Armenta A, Martin J, et al. Delayed diagnosis of post-surgical pyoderma gangrenosum: A multicenter case series and review of the literature. *International Journal of Surgery Case Reports.* 44 (2018), 152-156.
2. Tolkachjov SN, et al. Postoperative pyoderma gangrenosum (PG): the Mayo Clinic experience of 20 years from 1994 through 2014. J. Am. Acad. Dermatol., 73 (4) (2015), pp. 615-622
3. Monari P et al. Epidemiology of pyoderma gangrenosum: Results from an Italian prospective multicentre study. Int Wound J. 2018 Dec; 15(6): 875-879.
4. Tolkachjov SN et al. Postoperative Pyoderma Gangrenosum: A Clinical Review of Published Cases. Mayo Clin Proc. 2016 Sep; 91(9): 1267-79.
5. Ogata K et al. Pyoderma gangrenosum in an abdominal surgical site: a case report. Surg Case Rep. 2015 Dec; 1:122.
6. Wang C et al. Immune dysregulation in myelodysplastic syndrome: Clinical features, pathogenesis, and therapeutic strategies. Crit Rev Oncol Hematol. 2018 Feb; 122. 123-132.
7. Crowson AN, Mihm MC Jr, Magro C. Pyoderma gangrenosum: a review. J Cutan Pathol. 2003 Feb; 30(2): 97-107.
8. Le Cleach L et al. Is topical monotherapy effective for localized pyoderma gangrenosum? Arch Dermatol. 2011; 147: 101-3.
9. Thomas et al. Clinical outcomes and response of patients applying topical therapy for pyoderma gangrenosum: A prospective cohort study. J Am Acad Dermatol. 2016 Nov; 75(5): 940-949.
10. Kechichian E et al. Pediatric pyoderma gangrenosum: a systemic review and update. Int J Dermatol. 2017 May; 56(5): 486-495.
11. Leiphart PA et al. Suppression of pathergy in pyoderma gangrenosum with infliximab allowing for successful tendon depridement. JAAD Case Rep 2017 Dec 20; 4(1): 98-100.
12. Niamtu J 3rd. Pyoderma Gangrenosum After Facelift and Otoplasty Surgery: Case Presentations and Literature Review. J Oral Maxillofac Surg. 2018 Oct [Epub ahead of print].