Pyoderma gangrenosum (PG) is a rare inflammatory and ulcerative disorder of the skin. Despite the name, PG is not an infectious process. It is instead a neutrophilic dermatosis characterized by neutrophil-predominant cutaneous infiltrates. It is manifested clinically as an inflammatory papule or pustule that progresses to a painful ulcer with a well-defined erythematous to violaceous border and a purulent base. Non-specific laboratory findings include leukocytosis, elevated erythrocyte sedimentation rate, and elevated C-reactive protein. Fever is often present in this disorder. A retrospective study looking at the medical records of patients with PG found that pathergy was documented in 31% of cases. The term pathergy refers to the induction or exacerbation of PG in sites of incidental or iatrogenic trauma. Because clinical and laboratory findings in PG are nonspecific, diagnosis depends on ruling out other disorders that are manifested with cutaneous ulcerations. After the exclusion of other disorders, the management of PG usually involves topical corticosteroids or immunosuppressants for mild, localized cases and systemic corticosteroids or cyclosporine for severe, extensive cases. Because of the phenomenon of pathergy, surgical procedures should generally be avoided in PG. We present a case of PG that was initially misdiagnosed as an infectious olecranon bursitis and treated as such, leading to clinical deterioration of the patient until the correct diagnosis was made. The purpose of this report is to make readers aware of the presentation of PG, of its overlapping features with olecranon bursitis, and how misdiagnosis and mismanagement can lead to dramatic clinical deterioration.

Case report

A 49-year-old African American woman with a history of poorly controlled type 2 diabetes presented to the emergency department with swelling and pain over the right olecranon. Associated symptoms included subjective fever, chills, nausea, and vomiting. On presentation, the patient was afebrile and had a leukocytosis of 14,000/μL. The patient was treated with oral clindamycin and acetaminophen for suspected cellulitis. Three days later, the patient returned with increased elbow and forearm pain. She was again afebrile but this time had a leukocytosis of 21,000/μL. Erythrocyte sedimentation rate (ESR) was found to be 100 mm/h (normal, 0-20 mm/h), and C-reactive protein (CRP) level was 18.6 mg/dL (normal, 0.0-0.3 mg/dL). At this time, the emergency department and medical physicians diagnosed septic olecranon bursitis and cellulitis, and the patient was admitted to the Internal Medicine service. An emergency department swab wound culture from the olecranon bursa revealed the presence of methicillin-resistant Staphylococcus aureus, and empirical cefepime and vancomycin antibiotic therapy was initiated. During the next several days, the patient’s pain, swelling, and erythema around the elbow increased. In addition, the white blood cell (WBC) count remained elevated despite antibiotic treatment.

The orthopedic service was consulted at this point. On clinical examination, there was significant pain and swelling in the elbow and forearm with tenderness to palpation and pain with movement. The skin over the
Olecranon appeared friable with draining pustules. In addition, the patient appeared very ill. The patient was taken urgently for incision and débridement (I&D) of the right elbow. Medial and lateral skin incisions were made at the elbow to avoid further damage to the tenuous skin over the proximal ulna. Operative findings at this time were underwhelming except for significant watery tissue edema. Two antibiotic-impregnated cement rods were left under the skin. On postoperative day 1, a second orthopedic surgeon on "weekend rounds" evaluated the patient and found gross edema and spreading desquamation. The patient was immediately taken to the operating room for repeated I&D and extensile fasciectomy (Fig. 1, A and B) secondary to concern for necrotizing fasciitis. The patient was started on broad-spectrum antibiotics after the operation. Despite operative intervention, the patient's condition continued to deteriorate. She began spiking fevers, and her WBC count continued to climb (reaching >45,000/μL). Her extremity appeared worsened, with desquamation and drainage extending to the palmar hand (Fig. 1, C and D). Intraoperative blood and fluid cultures repeatedly came back negative. The infectious disease service was also consulted, and they recommended antibiotic treatment with vancomycin, clindamycin, and levofloxacin. On postoperative day 9, a third I&D of the elbow and forearm was performed. However, the patient's ulcers and leukocytosis failed to resolve. The possibility of a fungal infection was entertained after wound culture grew *Candida* species. The patient was started on a 10-day course of fluconazole. During this time, the WBC count reached a high of 46,000/μL. At this point, the orthopedic team recommended a dermatology consultation. Given the patient's history of present illness and hospital course, the dermatology consultant immediately suspected pyoderma gangrenosum (PG) as the most likely diagnosis. Because PG is a diagnosis of exclusion, biopsy of the ulcer was performed to...
rule out infection. Tissue culture came back negative for organisms, and biopsy results were nonspecific. Because neutrophilic infiltration was not the finding on biopsy, the primary medicine team doubted PG as the definite diagnosis. The patient was taken to the operating room on postoperative day 10 for a fourth I&D and placement of a negative-pressure wound vacuum. Despite the numerous interventions, the patient’s condition failed to improve during the next couple of weeks. The infectious disease service recommended stopping antibiotics at this point, given the patient’s continued fevers and leukocytosis despite 5 weeks of treatment with different antibiotics (vancomycin, cefepime, clindamycin, levofloxacin, and ceftriaxone). A few days later, the patient developed a lesion on her abdomen, very similar to the presenting elbow lesion. Biopsy of this lesion revealed neutrophilic dermatitis, consistent with PG.

The patient was immediately started on high-dose oral prednisone. In response, the patient’s skin lesions regressed both in the forearm and in the abdominal region. Her fever ceased, and her WBC count reached 12,200/μL after 1 week. Overall, the patient had a 53-day hospital course and showed improvement only after prednisone therapy in the last 10 days. Other complications during her prolonged hospital course included anemia, acute kidney injury, and an episode of cardiac arrest with pulseless electrical activity. After discharge from the hospital, the patient required 1 month of inpatient rehabilitation for general deconditioning and wound care. She received high-dose prednisone therapy for 2 full months before slowly being tapered down during the following 2 months. All wounds eventually healed. However, significant stiffness and deficits in range of motion remained in the affected extremity (Fig. 2).

Discussion

Septic olecranon bursitis typically is manifested with pain, erythema, and warmth around the elbow joint. In addition, fever, leukocytosis, and neutrophilia with bandemia are common findings in patients with this condition. ESR and CRP are also usually elevated.9,24 All of these characteristics were evident in our patient. Similar to what was initially believed to be the cause of our patient’s condition (cellulitis), septic olecranon bursitis is most frequently caused by contiguous spread of infection from cellulitis to the bursa.26,28 Treatment of septic olecranon bursitis includes antibiotic administration and drainage of infected bursal fluid.1,11 Antibiotic treatment is based on culture and susceptibility results. Severe infection, especially in patients with immunosuppressive comorbidities, requires hospital admission for intravenous administration of antibiotics.11 Continuous drainage of the infected bursa (by repeated needle aspiration or surgical drainage) is recommended in cases that require hospitalization.28

In a retrospective study looking at 343 cases of severe septic bursitis, 85% of patients achieved cure after 13 days of antibiotic treatment (surgery was also performed on the majority of the patients in the study).16 In contrast, our patient was treated with antibiotics and surgical débridement for more than 30 days with no clinical improvement.

Another diagnosis that was considered for our patient was necrotizing fasciitis. Necrotizing fasciitis is manifested clinically as localized erythema, swelling, calor, and tenderness that progress to skin breakdown and cutaneous gangrene over several days.10 Necrotizing fasciitis has a mortality rate of 24%.14 Diabetes (present in our patient) is one of the most important risk factors for necrotizing fasciitis, particularly type 1 (polymicrobial) necrotizing fasciitis.9 The diagnosis of this disorder is established surgically by visualizing fascial planes and muscle tissue in the operating room. Furthermore, surgical exploration allows early débridement and acqurement of tissue for culture.23 Surgical exploration and intervention should never be delayed while awaiting results of blood cultures, radiographic imaging, or skin aspirates.25 The dramatic worsening of our patient’s condition after the initial débridement resulted in a second orthopedic surgeon fearing the presence of necrotizing fasciitis. However, fasciectomy and repeated radical débridement only worsened her condition.

The term *pyoderma gangrenosum* was first coined by Brunsting et al in 1930 to describe an ulcerative cutaneous disorder that they believed was caused by bacterial infection.1 In 1957, Percival studied the histopathologic characteristics of the primary lesion in PG and concluded that it is a neutrophilic dermatosis rather than an infectious process.15 Nonetheless, the original name has persisted. Clinically, an inflammatory papule, pustule, vesicle, or nodule initially forms that then expands into an ulcerative erosion.7,17 Nonspecific findings commonly seen in PG include fever, leukocytosis, and elevated ESR and CRP.21,27 Our patient had all of these findings.

Although the exact etiology is unknown, it is believed that abnormal immunologic reactions play a role in PG.13,21 PG can be classified into 4 types: ulcerative (classic), pustular, bullous (atypical), and vegetative.19,22 Ulcerative is the most common and bullous is the least common type.12,19,22 More than 50% of patients with PG have an associated disorder, such as inflammatory bowel disease, arthritis, and hematologic disorders (i.e., myelogenous leukemia, hairy cell leukemia, myelofibrosis, and monoclonal gammopathy).12,13,21 On the other hand, 40% to 50% of cases involve only the skin and are referred to as idiopathic PG.21 Our patient most likely suffered from idiopathic PG, given that subsequent evaluation did not reveal any associated disorders.

For diagnosis of PG, the first step is to rule out disorders that are manifested similarly (infectious diseases, malignant disease, vasculitis, insect bites, vascular insufficiency, and factitious ulcerations) by culturing exudates and obtaining tissue biopsy specimens.7,21 Pathergy (induction or exacerbation of ulcers in PG due to incidental or iatrogenic trauma) may result even from small procedures, such as a biopsy. Nevertheless, biopsy should be performed to rule out malignant disease,
vasculitis, and infection.\textsuperscript{7,21} In our patient, pathergy played a large role in the continued misdiagnosis and deterioration as each surgery resulted in a worse-appearing limb. The second step in diagnosis is to determine whether any of the associated disorders (such as ulcerative colitis, arthritis, and hematologic disorders) are present.\textsuperscript{7,21} The dermatology team carried out these steps and efficiently made the correct diagnosis in our patient. Establishment of the correct diagnosis is vital before management is started because certain therapies used for PG can be ineffective or harmful in other clinically similar diseases (and vice versa).

The severity of the disorder influences the approach to treatment of PG.\textsuperscript{7,21} In mild, localized disease, initial treatment usually involves high-potency or superpotent topical corticosteroid or tacrolimus. For patients with more extensive disease, oral glucocorticoids are indicated. Alternatively, cyclosporine, with or without systemic glucocorticoids, may be used as first-line therapy.\textsuperscript{20,21}

Surgery on the affected area is generally avoided in PG because of the phenomenon of pathergy.\textsuperscript{21} Surgical procedures may be considered in cases in which accumulation of necrotic tissue presents a risk for infection or when tendons or ligaments are exposed in the ulcer bed.\textsuperscript{6,20} An important caveat is to limit surgical procedures to periods of good disease control under systemic immunosuppressive therapy to minimize the likelihood of pathergy.\textsuperscript{7}

After appropriate therapy for PG, signs of clinical improvement may be evident within days. However, complete ulcer healing often takes weeks to months.\textsuperscript{17} Our patient had extensive disease yet began to show signs of improvement of her ulcers after a few days of oral prednisone therapy. Her WBC count went from 25,300/\text{mL} to 12,200/\text{mL} after just 6 days of therapy. The prednisone dose was slowly tapered after 2 months for a total of 4 months of therapy. In addition, she was prescribed alendronate, vitamin D, and calcium to protect against glucocorticoid-induced osteoporosis.

Overall, PG is more commonly found on the lower extremity and trunk.\textsuperscript{13,18} Given the rarity of PG in the upper extremity, little has been published in the upper extremity surgery literature about this topic.\textsuperscript{27} One case series in 2001 described only 7 patients with PG involving the hand during a 7-year period from 2 different institutions. All the patients in that case study had an associated systemic disease, and all were initially misdiagnosed as having an infection.\textsuperscript{12} Choe et al reported an isolated case of PG involving the wrist.\textsuperscript{8} Bullous PG (the least common type of PG) is more likely

\textbf{Figure 2}  (A-E) At a follow-up visit 2 months after discharge from the hospital, healed PG ulcers can be seen. (A) The patient had stiffness and limited elbow range of motion (the patient is attempting maximum elbow flexion in this picture). (C) The black arrow shows superficial forearm PG ulcer; the white arrow shows superficial abdominal ulcer, biopsy of which led to the correct diagnosis. Ulcers show improvement compared with when the patient was in the hospital. (E) Healed palmar and forearm PG lesions after 2 months of prednisone therapy.
to occur on the arms and face than on the legs.\textsuperscript{19,21,22} However, even with this type, a review of the literature hints that elbow involvement is even rarer than hand and wrist involvement. We were able to find only 1 case reported in the literature of PG presenting on the elbow. The patient in this case had a history of chronic polyarthritis and also had a PG ulcer on her neck.\textsuperscript{12} To our knowledge, there are no other reported cases of PG presenting on the elbow in the literature.

Conclusion

The case presented highlights how easily PG can be misdiagnosed and mistreated. The rarity, nonspecificity of findings, and diversity of the pathogenesis of this disorder result in the diagnostic challenge. Nonetheless, to manage this disorder properly and to obtain a positive outcome, an accurate diagnosis is vital. Evidently, surgical procedures and antibiotics (effective methods of treatment in infectious diseases such as septic olecranon bursitis) were ineffective and perhaps harmful in this case. Corticosteroid therapy, on the other hand, resulted in clinical improvement. Therefore, whenever a patient presents with ulcerative cutaneous lesions that resemble an infectious process such as olecranon bursitis, PG must be considered in the differential diagnosis.

Disclaimer

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