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A Case Report of Ecthyma Gangrenosum affecting the Thumb

Corey T. Clyde, MD[1]
John M. Roberts, MD[1]
John R. Lien, MD[2]
Benjamin L. Gray, MD MSCE[1]

[1]Department of Orthopaedic Surgery, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA

[2]Department of Orthopaedic Surgery, University of Michigan, Ann Arbor, MI

Introduction

Ecthyma gangrenosum is an atypical cutaneous vasculitis classically associated with immunocompromised or critically ill hosts. [1-2] This particular vasculitis results from bacteremia or direct inoculation of an infecting microorganism, commonly the pathogen Pseudomonas aeruginosa.[3-4] Most often lesions appear in the gluteal region, perineum, or axilla; occasionally affecting the extremities.[5-6] Appearance of the lesions on the hand and digits are particularly rare, and we present such a case diagnosed and treated at our center.

Case Information

A 33-year-old right hand dominant female presented to the emergency department with the chief complaint of a right thumb blister. The patient reported a 2-day history of an isolated right thumb lesion with accompanying erythema, swelling, and warmth. The patient noted worsening symptoms with the blister developing into an open wound at the radial border of the right thumb interphalangeal joint (Figure 1). Otherwise, she did not report systemic complaints.

Regarding pertinent past medical history, the patient did have a previous diagnosis of Sjogren's disease along with low grade B cell marginal zone lymphoma. She had recently completed her last dose of a bi-monthly rituximab maintenance regimen approximately one month prior to presentation. She was considered to be in remission by her hematologist.

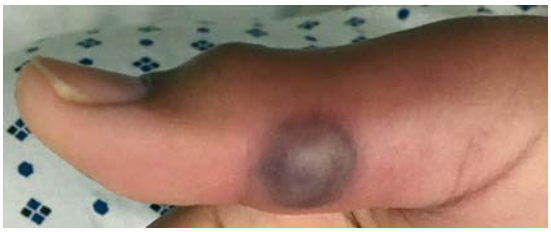

Figure 1. Lesion at time of initial presentation.

On examination her vital signs were stable. The patient was noted to have an open wound overlying the radial border of the interphalangeal joint of the right thumb measuring 1.5cm by 1.3cm surrounded by a gray border, which was necrotic in appearance. Radiographs taken of the right hand appeared within normal limits without acute or chronic osseous changes. An acute inflammatory lab panel was obtained revealing white blood cell count 5.7 K/μL, C-reactive protein 139.6 mg/L, erythrocyte sedimentation rate 87 mm/h.

Initial assessment included a differential diagnosis of abscess, interphalangeal joint septic arthritis, and flexor tenosynovitis. The patient underwent a bedside right thumb irrigation and non-excisional debridement. She was admitted to the hematology/oncology service given her history, and was started on a broad-spectrum antibiotic regimen including vancomycin, piperacillin/tazobactam, ampicillin/sulbactam, metronidazole, and cefepime. Infectious disease, dermatology, and wound care consults were also obtained.

There was a lack of improvement in the clinical appearance of the right thumb after the index procedure. Two days later the patient was taken to the operating room for more formal debridement (Figure 2a and 2b). A right thumb interphalangeal joint arthrotomy along with irrigation and excisional debridement was performed. The right thumb was incised in a mid-axial fashion along the radial border. Non-viable tissue including skin, subcutaneous fat, and fascia were excised sharply. The neurovascular

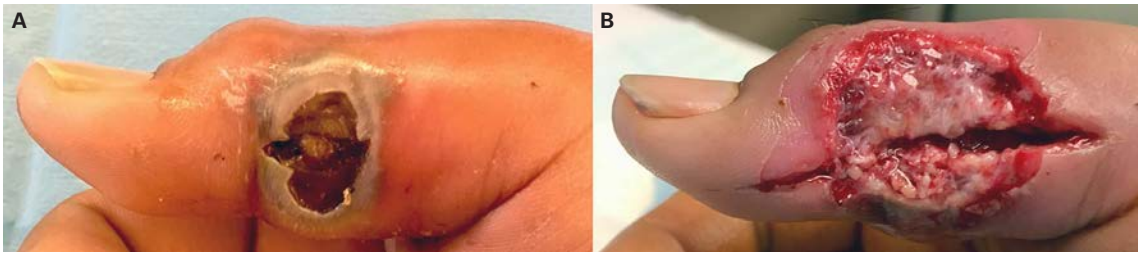

Figure 2 (A) Appearance prior to formal surgical procedure; **(B)** Appearance post-operative day 2.

bundle was isolated and protected throughout the case. Dissection was carried volarly and dorsally. In the dorsal area of the thumb a collection of murky fluid and necrotic fat was encountered. The flexor tendon sheath did not appear involved. The thumb interphalangeal joint contained murky fluid and synovitis. Four microbiology specimens and one pathology specimen were obtained. The wounds were thoroughly irrigated with normal saline. The patient tolerated the procedure well without complications.

Ultimately, the intra-operative cultures returned with growth of Pseudomonas aeruginosa. Blood cultures remained without growth. Other testing performed, which was all negative included VZV, HSV, AFV, mycobacteria, and fungal studies. At the recommendation of the infectious disease team, the patient's broad spectrum antibiotic regimen was narrowed to levofloxacin 750 mg daily for a 14-day course. Pathology analysis of the excised skin demonstrated parakeratosis, dermal necrosis, and acute inflammation of the subcutaneous adipose tissue with numerous bacilli present within dermis, consistent with ecthyma gangrenosum.

The patient's subsequent hospital course included gradual improvement in the appearance of the thumb. During the post-operative period she was noted to be neutropenic with white blood cell count 2.4 K/μL and absolute neutrophil count 266 /mm^3. The primary team began filgrastim 300 mcg to stimulate her neutrophil count**.** The total hospital course was 7 days, with the patient being discharged to home in stable condition.

In the subsequent weeks the patient was followed closely. She continued to demonstrate clinical improvement without setbacks. Her last orthopaedic hand surgery follow-up was approximately four months post-operatively at which point her thumb wound was healed (Figure 3).

Prior Reports and Relevant Literature

Ecthyma gangrenosum was first used as a diagnosis by Hitschmann and Kreibich in 1897.[5] There have since been chapters, case series, and case reports describing the condition. The association with immunocompromised patients is well known, with few reports of infections in immunocompetent individuals.[5, 7-9] The characteristic lesion begins as a macule, vesicle, or bulla; then progresses to an indurated ulcer. The ulcer sloughs to form a gangrenous ulcer with necrotic center, black eschar, and erythematous ring.[6] The maturation of the lesion is typically rapid, occurring within 12 to 18 hours.

Histologically, the lesion results from perivascular bacterial invasion of the media and adventitia layers of small vessels by gram-negative bacilli with secondary ischemic necrosis.[9] Exotoxins produced by bacteria mediate local tissue degradation. The aforementioned vasculitic changes may be seen by histologic examination of pathology specimens.[10-11] Pseudomonas aeruginosa is the most common pathogen, but Staphylococcus aureus, Citrobacter freundii, Morganella morganii, Candida species, Aeromonas hydrophila, Seratia marcescens, and other species have been reported.[12-14]

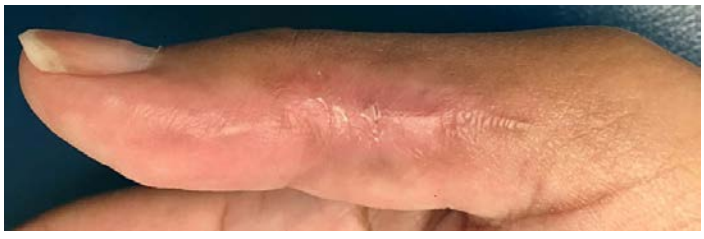

Figure 3. Appearance at four months follow-up.

Prompt recognition, diagnosis and evaluation of this process including blood cultures, wound cultures, and tissue biopsy are critical to improving prognosis and directing clinical decision making. Treatment involves the use of empiric antimicrobial therapy with anti-pseudomonal activity for a lesion suspicious for ecthyma gangrenosum. Excisional debridement may also be of benefit for more aggressive lesions.[14]

Koumaki at el. reported on a 47-year-old male diagnosed with ecthyma gangrenosum on the dorsum of the right hand.[15] The patient had been diagnosed with acute myeloid leukemia receiving first, second, and third line chemotherapy treatments. The hand lesion developed in the setting of acute bacteremia. Wound and blood cultures grew Klebsiella pneumoniae and Streptococcus vestibularis. A pathology specimen revealed vascular necrosis with many surrounding bacteria consistent with a diagnosis of ecthyma gangrenosum. The patient underwent surgical debridement and received antibiotic treatment, ultimately with complete resolution of the lesion three months after onset.[15]

Aygencel et al. reported on an 80-year-old male with a lesion on the dorsum of the left hand diagnosed as an ecthyma gangrenosum-like eruption.[16] In this case, the patient had a medical history significant for cardiovascular disease, hypertension, chronic obstructive pulmonary disease, and chronic kidney disease. He was admitted to the medical intensive care unit in critical condition with a new diagnosis of multiple myeloma. The lesion presented on the hand several weeks into the admission. Wound and blood cultures grew Burkholderia cepacian, which was antibiotic resistant according to the sensitivity testing performed. The patient ultimately died during the hospitalization from sepsis.[16]

Discussion

To our knowledge there are no other descriptions of ecthyma gangrenosum in the orthopaedic hand surgery literature. Of the available case series and case reports pertaining to this diagnosis published in other specialty journals there are two patients with ecthyma gangrenosum affecting the hand, none with digital lesions. For those two patients, both were immunocompromised, acutely ill, had an isolated dorsal hand lesion, neither grew the typical Pseudomonas aeruginosa as the infecting organism.

Our patient represents a unique instance of ecthyma gangrenosum affecting the thumb in isolation. Although our patient too was an immunocompromised host, she was not acutely ill at the time of presentation. Otherwise, the lesion presented with a history, physical examination, microbiology, and pathology characteristic for the diagnosis of ecthyma

gangrenosum. She was successfully treated with a combination of antibiotics and surgical interventions.

Conclusions

Ecthyma gangrenosum is a rare infective vasculitis that may carry a poor prognosis for immunocompromised patients. The lesions may affect the hand and digits. The consulting hand surgeon should keep ecthyma gangrenosum in the differential diagnosis during evaluation of hand lesions in immunocompromised hosts, particularly those lesions that fail to respond to conventional treatment. Biopsies are essential to target the antibiotic regimen, and histological analysis of skin and dermis specimens can help confirm the diagnosis and guide treatment.

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