

Pyoderma Gangrenosum and Extensive Aseptic Chest Wall Abscess in a Patient with Inflammatory Bowel Disease

Igor Snast MD¹, Iris Ostfeld MD², Lev Pavlovsky MD PhD¹, Emilia Hodak MD^{1,4} and Anat Gafter-Gvili MD^{2,3,4}

Departments of ¹Dermatology and ²Medicine A, Rabin Medical Center (Beilinson Campus), Petah Tikva, Israel

³Institute of Hematology, Rabin Medical Center (Beilinson Campus), Petah Tikva, Israel

⁴Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

KEY WORDS: aseptic abscess, inflammatory bowel disease (IBD), neutrophilic dermatosis, neutrophils, pyoderma gangrenosum (PG)

IMAJ 2018; 20: 712–713

P pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis, which presents as a rapidly expanding ulcer with a necrotic undermined border. More than 50% of patients with PG have an antecedent, coincident, or subsequent associated systemic disease, most commonly inflammatory bowel disease (IBD), arthritis, and hematologic disease (i.e., acute and chronic myelogenous leukemia) [1]. Similarly, aseptic abscess (AA) is an inflammatory condition characterized by deep sterile collections of neutrophils, clinically mimicking bacterial abscess [2]. We describe a patient with IBD who presented with a unique combination of an extensive chest wall AA concomitantly with necrotic ulcers characteristic of PG.

PATIENT DESCRIPTION

A 24 year old woman with newly diagnosed IBD, presented with a firm, tender, erythematous nodule located on the proximal clavicle, with sternoclavicular joint pain. At that time, she was being treated with 0.5 mg/kg prednisone and vedolizumab after failing to improve with infliximab, mesalazine, and low-dose prednisone. A chest

computed tomography (CT) scan depicted a large subcutaneous chest wall abscess extending within the pectoralis muscle with sternal destruction. An incision was performed with drainage of purulent fluid. Broad-spectrum antibiotics were initiated. Near the chest wound, two satellite rapidly enlarging ulcers with undermined violaceous borders appeared.

Due to progressive chest wall necrosis on a consecutive CT, radical debridement with excision of pectoralis major muscle was performed, resulting in rib exposure [Figure 1A]. On the thigh, an erythematous nodule appeared, which rapidly underwent necrotic changes, leading to a painful ulcer with violaceous-rolled border [Figure 1B]. Skin biopsies taken from the ulcer base on

the thigh and chest wound revealed a diffuse dermal infiltrate of neutrophils extending into chest wall striated muscles, with focal fibrinoid necrosis of blood vessel walls.

Laboratory evaluation demonstrated elevated inflammatory markers; negative serologies for hepatitis B/C, human immunodeficiency virus, cytomegalovirus, and atypical bacteria; negative Mantoux and QuantiFERON tests; negative blood, skin, and purulent fluid cultures including on enrichment media; negative pan-bacterial; and pan-fungal blood polymerase chain reaction.

A diagnosis of chest wall AA with characteristic thigh and chest PG lesions was made based on the clinical presentation, histologic findings, and absence of

Figure 1. [A] Large surgical wound after subcutaneous abscess incision and drainage extensive enough to expose periosteum and muscle, with two inferior satellite ulcers [B] Close up view of pyoderma gangrenosum of left thigh



infectious pathogens [1]. Intravenous pulse methylprednisolone was initiated on 4 consecutive days, followed by oral 1.3 mg/kg prednisone with a concomitant cyclosporine 5 mg/kg. Given the lack of an adequate clinical response and elevation of inflammatory markers when steroid tapering was attempted, intravenous immunoglobulins 3 gr/kg were added. Over 3 weeks, the thigh ulcer healed completely and the chest necrolysis ceased with granulation tissue filling wound-bed. Two weeks later, the patient underwent reconstructive surgery. Exposed ribs were covered with a muscular flap of the remaining pectoralis major so that the entire wound was covered with a split-thickness skin. Prednisone and cyclosporine were slowly tapered. Subsequently, adalimumab was initiated due to IBD exacerbation without skin lesion recurrence.

COMMENT

AA represents a new clinical entity characterized by deep neutrophilic abscesses,

most commonly of intra-abdominal origin, negative infectious workup, lack of a clinical response to antibiotics, and a dramatic response to steroids [2,3]. Excluding the latter, which could not be assessed as AA was surgically treated, our patient met all of the criteria. Similar to our patient, in the largest cohort to date of 30 patients with AAs, most patients (n=21) had IBD followed by neutrophilic dermatosis (n=6), and relapsing polychondritis (n=3) [2].

Although most cases of AA were shown to involve the spleen. Our case demonstrates that AAs can potentially appear anywhere in the body. Our patient presented with the typical appearance of PG, another rare dermatosis mediated by neutrophils, concurrently with AA. The overlap between the two conditions was also evident in a recent literature review of IBD-associated AA, in which 2 of 37 patients (5.5%) presented with both disorders [3]. This finding suggests that common immunologic pathways may mediate both disorders [4].

Given the rarity of AA and the lack of clear diagnostic criteria, the correct diagnosis and appropriate treatments are frequently delayed. Greater knowledge of AA among physicians will promote early diagnosis and effective treatment.

Correspondence

Dr. I. Snast

Dept. of Dermatology, Rabin Medical Center (Beilinson Campus), Petah Tikva 4941492, Israel
Phone: (972-3) 937-6653
email: snastigor@gmail.com

References

1. Ahronowitz I, Harp J, Shinkai K. Etiology and management of pyoderma gangrenosum: a comprehensive review. *Am J Clin Dermatol* 2012; 13 (3): 191-211.
2. André MFJ, Piette J-C, Kémény J-L, et al. Aseptic abscesses: a study of 30 patients with or without inflammatory bowel disease and review of the literature. *Medicine (Baltimore)* 2007; 86 (3): 145-61.
3. Bollegala N, Khan R, Scaffidi MA, et al. Aseptic abscesses and inflammatory bowel disease: two cases and review of literature. *Can J Gastroenterol Hepatol* 2017; 2017: e5124354.
4. Butler D, Shinkai K. What do autoinflammatory syndromes teach about common cutaneous diseases such as pyoderma gangrenosum? A commentary. *Dermatol Clin* 2013; 31 (3): 427-35.

Capsule

Gut check for a promising HIV treatment

Eradicating human immunodeficiency virus (HIV) in infected patients likely requires disrupting the reservoir of infected T cells in the gastrointestinal tract. One approach may be targeting cells expressing the integrin $\alpha 4\beta 7$, which has been tested in simian immunodeficiency virus models and is an approved therapy for inflammatory bowel disease. **Uzzan** and colleagues studied a small cohort of HIV-infected individuals on antiretroviral therapy who began receiving an antibody

against $\alpha 4\beta 7$ as a treatment for their mild inflammatory bowel disease. Longitudinal colonoscopies revealed that the anti- $\alpha 4\beta 7$ therapy disrupted local lymphoid aggregates. The treatment was well tolerated, but long-term effects on the HIV reservoir remain to be determined.

Sci Transl Med 2018; 10: eaau4711
 Eitan Israeli

Capsule

Gene editing and muscular dystrophy

Duchenne muscular dystrophy (DMD) is characterized by progressive muscle weakness and a shortened life span. The disease is caused by mutations that reduce or prevent expression of dystrophin, an essential structural protein in skeletal and heart muscle. The gene editing technology CRISPR-Cas9 can correct disease-causing mutations and has yielded promising results in mouse models of DMD. In a small, short-term study, **Amosii** et al. tested this strategy in a dog

model of DMD that exhibits many features of the human disease. Intramuscular or systemic delivery of the gene editing components resulted in a substantial increase in dystrophin protein levels in skeletal and heart muscle. Restoration of dystrophin expression was accompanied by improved muscle histology.

Science 2018; 362: 86
 Eitan Israeli