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PYODERMA GANGRENOSUM PRESENTING AS FOURNIER'S GANGRENE

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ABSTRACT

We report a case of pyoderma gangrenosum presenting as Fournier's gangrene. Although both processes have a similar presentation effective management is markedly different. Whereas broad-spectrum antibiotics and aggressive surgical débridement are necessary to control Fournier's gangrene, immediate institution of corticosteroids and local wound care are indicated for pyoderma gangrenosum. (*J. Urol., 144: 984–986, 1990*)

Pyoderma gangrenosum is a rare, noninfectious, ulcerative disease of the skin.[1–4] The most commonly affected sites are the lower extremities and, rarely, the upper torso and face. Only 2 cases affecting the male genitalia have been reported previously.[5,6] Approximately 50% of the patients will have an associated systemic disorder, inflammatory bowel disease being the most common.[7] The cause of pyoderma gangrenosum is unknown. Histological changes are not specific and laboratory findings are not pathognomonic, making the diagnosis one of exclusion.

Treatment with systemic and topical steroids is generally effective. Although pyoderma gangrenosum may present similarly to Fournier's gangrene, effective management is markedly different. Whereas broad-spectrum antibiotics and aggressive surgical débridement are necessary to control necrotizing infections,[8] immediate institution of corticosteroids and local wound care are indicated for pyoderma gangrenosum.

CASE REPORT

A 39-year-old man was admitted to a community hospital for fever and a progressive, purulent lesion of the penis. He had been treated for a penile discharge with procaine, penicillin and tetracycline for culture proved gonorrhea (penicillin-sensitive) 1 month previously. Other than mild mental retardation the medical history was unremarkable (no trauma, diabetes, vascular disease, or intravenous drug or alcohol abuse). At hospitalization the patient denied hematuria and dysuria, and was taking no medications. On physical examination the temperature was 103.2F, and the penis showed extensive purulence and necrosis of the shaft on the lateral and ventral surfaces. The scrotum and testicles were uninvolved, the rectum and perineum were normal, and there was no inguinal adenopathic condition. Laboratory data revealed a white blood count of 28 $\times 10^3/\mu$l. with a left shift. Multiple blood and urine cultures were negative. Tissue cultures from the penile lesion for Herpes simplex, acid-fast bacteria, gonorrhea, and aerobic and anaerobic bacteria also were negative. Serological testing for antideoxyribonucleic acid antibody and rheumatoid factor were negative. Chest x-rays were normal and plain abdominal films revealed no air in the soft tissue of the perineum.

Diagnostic skin biopsy performed the day after hospitalization revealed extensive inflammation without evidence of cause. Despite local wound care and intravenous antibiotics the patient continued to have fever and an elevated white blood count. Even after surgical débridement of the necrotic penile tissue the inflammatory process extended to the scrotum and suprapubic region. Débridement was repeated and broad-spectrum intravenous antibiotics were continued (fig. 1).

Diagnostic tests, including an abdominopelvic computerized tomography (CT) scan, echocardiogram and gallium scan, all failed to identify an occult source of infection. Six days after hospitalization the patient suffered a painful right hip lesion that progressed rapidly to a necrotic, purulent ulcer. Cultures of this lesion showed no bacterial growth. A similar lesion soon developed on the right shoulder as well as a group of pustules below the left eye. With continuing fever, elevated white blood count and progression of the penile lesion as well as new satellite lesions, the patient was transferred to our institution for further care. Working diagnosis was an inadequately débrided necrotizing soft tissue infection of the genitalia.

Examination revealed an extensively débrided penis with focal loss of tunical tissue around the corporeal bodies. The urethra was intact. The scrotum was markedly edematous and erythematous with multiple sites of purulent discharge. The suprapubic region was similarly involved.

The genitalia and perineum were extensively débrided and a suprapubic cystostomy tube was placed. The remaining necrotic tissue around the penile shaft was débrided, further exposing the cavernous bodies and corpus spongiosum, which had been separated by earlier surgical intervention (fig. 2). The satellite lesions also were extensively débrided.

Postoperatively, broad-spectrum antibiotics and wet-to-dry dressing changes were continued. Within 24 hours of débridement the cut edges of the wounds became necrotic and purulent, and close examination revealed new pustules with well defined erythematous borders. Despite repeated débridements (fig. 1), skin loss progressed and fever persisted. Blood and tissue cultures for aerobic and anaerobic bacteria as well as fungi continued to be negative, except for 1 tissue swab of Staphylococcus epidermidis.

Microscopic review of the tissue obtained during surgical débridement showed extensive ulceration and necrosis with massive infiltration by neutrophils (fig. 3). At the junction between the necrotic area and the adjacent skin the ulcer edge was markedly undermined by a dense infiltrate of neutrophils. A distinct vasculitis was not present nor was there evidence of bacteria or fungi.

The diagnosis of pyoderma gangrenosum was made because all microbiological cultures were negative, satellite lesions had developed, and the wound progressed despite surgical débridement and antibiotics. All antibiotics were stopped and a course of intravenous steroids (methylprednisolone sodium succinate at 60 mg. every 12 hours) was instituted. Within 24 hours the temperature normalized and the progressive skin necrosis stopped. The white blood count slowly returned to normal and with wet-to-dry dressing all wounds began to granulate.

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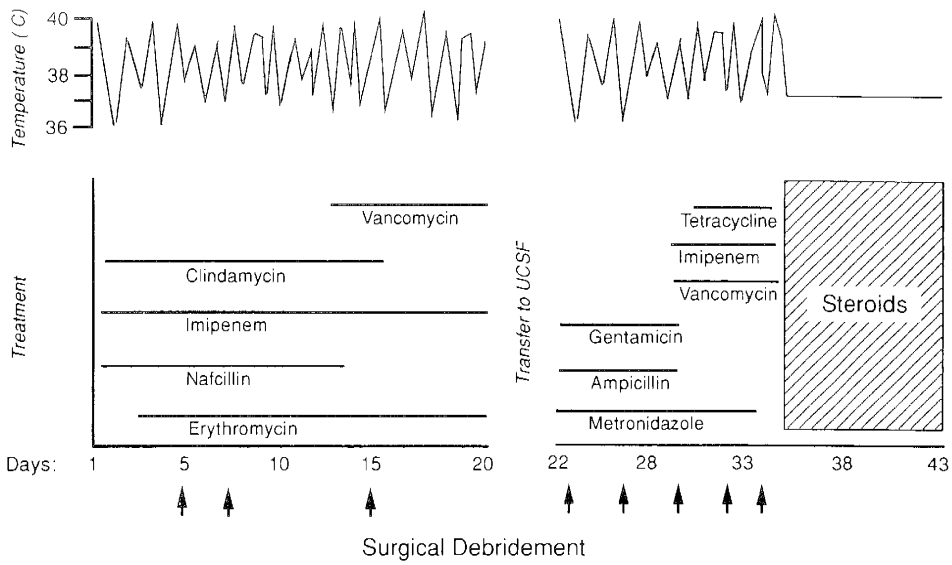

FIG. 1. Clinical course of patient with pyoderma gangrenosum that presented as Fournier's gangrene

FIG. 2. Extensive skin loss of penis and surrounding genitalia. Note separation of corpus spongiosum from cavernous bodies.

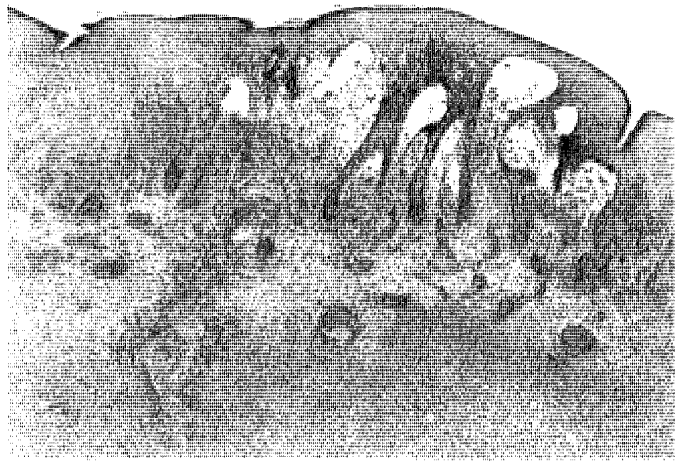

FIG. 3. Débrided skin in pyoderma gangrenosum. Note dense infiltration of neutrophils. Reduced from ×45.

Once the diagnosis of pyoderma gangrenosum was confirmed the patient was investigated for associated systemic disease. Normal findings on colonoscopy ruled out inflammatory bowel disease. Serum electrophoresis showed no associated monoclonal gammopathy. Complete blood count with T-cell analysis revealed no quantifiable immune defect and testing for the acquired immunodeficiency syndrome was negative.

During 3 weeks the steroid dosage was tapered to 10 mg. per day. Skin grafts were applied successfully to the right hip and shoulder wound. The testicles were initially implanted into the anterior thighs for wound management, with subsequent reconstruction of the scrotum and penile skin with split-thickness skin grafts and local skin flaps. Presently, the patient is taking low dose steroids every other day and there is no evidence of recurrent pyoderma gangrenosum.

DISCUSSION

Pyoderma gangrenosum was first described as an ulcerative skin disease in 1930 by Brunsting and associates.[4] The ulcers can be painful and many patients will have an associated high fever.[1,2] When the lesions characteristic of pyoderma gangrenosum are present the diagnosis is not difficult. These are typically sterile pustules that coalesce or break down to form necrotic ulcers,[1,2] which may be single or multiple and range from 1 to 30 cm. They are surrounded by an erythematous zone extending as an areola into the normal skin. However, in some patients lesions may be atypical with evolving pustules that resemble folliculitis or chemical dermatitis, such as iododerma or bromoderma. Pyoderma gangrenosum lesions may arise and regress spontaneously or they may occur at an area of trauma (pathergy) or surgical débridement.[9,10] Healing will occur by re-epithelialization once the inflammatory process has been interrupted.

Pyoderma gangrenosum has been classified as a neutrophilic dermatosis, which is reflected by the massive infiltration of polymorphic leukocytes histologically (fig. 3).[11] Subepidermal abscesses may be seen as well as acanthosis and pseudoepitheliomatous changes at the periphery of the ulcer. Granuloma formation and vasculitis are not present.

The cause of pyoderma gangrenosum is unknown. Numerous attempts to culture bacteria, fungi and viruses have failed consistently to reveal an infectious agent. An increased incidence of circulating immune complexes (IgA), anergy to various skin tests, and defective polymorphonuclear leukocyte chemo-

taxis and bacterial killing ability have been noted, suggesting an inherent immunological defect.[11-14] This may account for the cutaneous lesions.

At our institution we have had extensive experience in the treatment of patients with necrotizing soft-tissue infections of the perineum and genitalia (Fournier's gangrene).[8] During the last 15 years 29 patients have been treated for this often fatal disease (38% mortality in our group). Infections have tended to be polymicrobial, with an average of 4 different bacterial species cultured per patient (range 1 to 9). Typical presenting features include pain, swelling, fever and skin necrosis, as can be seen in pyoderma gangrenosum. However, satellite lesions do not occur in necrotizing infections, and the wound typically responds to surgical débridement and broad-spectrum antibiotics.

The accurate diagnosis of pyoderma gangrenosum is essential, since surgical débridement may lead to further tissue destruction (pathergy) and progression of the cutaneous lesions.[10] The differential diagnosis of an ulcerative cutaneous lesion involves infectious agents, halogenodermas, vasculitis, insect bites, arterial and venous insufficiency, factitious ulcerations and, as in this case, pyoderma gangrenosum. When the diagnosis is questionable histological analysis can be helpful. In this case negative cultures, satellite lesions, and the failure to respond to surgical débridement and broad-spectrum antibiotics are confirmatory (see table).

Once the diagnosis of pyoderma gangrenosum is suspected corticosteroids should be instituted. For smaller lesions injection of local steroids (triamcinolone at 30 mg./ml.) into the border may be effective. This therapy should be accompanied by local wound care with dressing changes, whirlpool therapy and silver sulfadiazine or hydrocolloid occlusive wound dressing. For refractory cases high doses of corticosteroids have been useful (1 gm. in 150 ml. dextrose in water given intravenously during 1 hour). Other effective agents include dapsone (G6PD screening necessary), sulfapyridine (also effective for inflammatory bowel disease) and, to a lesser extent or as an adjunct, azathioprine and clofazimine. The occasional patient who is unresponsive to steroids might benefit from the immunosuppressive agent cyclosporine A.[15]

In conclusion, although pyoderma gangrenosum can present as Fournier's gangrene, the treatment of this unusual ulcerative skin disease differs, and includes conservative local care and systemic and/or local steroids rather than surgical débridement and antibiotics.

Clinical characteristics distinguishing Fournier's gangrene from pyoderma gangrenosum

	Fournier's Gangrene	Pyoderma Gangrenosum
Pos. cultures	Yes	No
Fever	Yes	Yes
Pain	Yes	Yes
Skin necrosis	Yes	Yes
Satellite lesions	No	Yes
Response to steroids	No	Yes
Response to surgery and antibiotics	Yes	No

REFERENCES

1. Callen, J. P.: Pyoderma gangrenosum and related disorders. Med. Clin. N. Amer., **73:** 1247, 1989.
2. Prystowsky, J. H., Kahn, S. N. and Lazarus, G: Present status of pyoderma gangrenosum: review of 21 cases. Arch. Dermatol., **125:** 57, 1989.
3. Malkinson, F. D.: Pyoderma gangrenosum vs malignant pyoderma. Lumpers vs splitters. Editorial. Arch. Dermatol., **123:** 333, 1987.
4. Brunsting, L. A., Goeckerman, W. H. and O'Leary, P. A.: Pyoderma (ecthyma) gangrenosum. Clinical and experimental observations in five cases occurring in adults. Arch. Dermatol. Syph., **22:** 655, 1930.
5. Harto, A., Gutiérrez Sanz-Gadea, C., Vives, R., Romero Maroto, J. and Ledo, A.: Pioderma gangrenoso en pene. Actas Urol. Esp., **9:** 263, 1985.
6. Sanusi, I. D., Gonzalez, E. and Venable, D. D.: Pyoderma gangrenosum of penile and scrotal skin. J. Urol., **127:** 547, 1982.
7. Thornton, J. R., Teague, R. H., Low-Beer, T. S. and Read, A. E.: Pyoderma gangrenosum and ulcerative colitis. Gut, **21:** 247, 1980.
8. Baskin, L. S., Carroll, P. R., Cattolica, E. V. and McAninch, J. W.: Necrotizing soft-tissue infections of the perineum and genitalia: bacteriology, treatment and risk assessment. Brit. J. Urol., **65:** 524, 1990.
9. Work, B. A., Jr.: Pyoderma gangrenosum of the perineum. Obst. Gynec., **55:** 126, 1980.
10. Fulbright, R. K., Wolf, J. E. and Tschen, J. A.: Pyoderma gangrenosum at surgery sites. J. Dermatol. Surg. Oncol., **11:** 883, 1985.
11. Bentley-Phillips, C. B., Cooper, R. C. and Hallet, A. F.: Pharmacological modulation of neutrophil phagocytic function in a patient with recurrent sepsis, pyoderma gangrenosum and impaired phagocytosis. Brit. J. Dermatol., **106:** 687, 1982.
12. Delescluse, J., De Bast, C. and Achten, G.: Pyoderma gangrenosum with altered cellular immunity and dermonecrotic factor. Brit. J. Dermatol., **87:** 529, 1972.
13. Lazarus, G. S., Goldsmith, L. A., Rocklin, R. E., Pinals, R. S., de Buisseret, J.-P., David, J. R. and Draper, W.: Pyoderma gangrenosum: altered delayed hypersensitivity and polyarthritis. Arch. Dermatol., **105:** 46, 1972.
14. Aram, H.: Malignant pyoderma. Report of a case associated with a disorder of the immune system. Dermatologica, **171:** 112, 1985.
15. Curley, R. K., Macfarlane, A. W. and Vickers, C. F. H.: Pyoderma gangrenosum treated with cyclosporin A. Brit. J. Dermatol., **113:** 601, 1985.