Pyoderma Gangrenosum: A Case Series

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Pyoderma gangrenosum is a rare, ulcerative, non-infectious neutrophilic dermatosis, commonly associated with underlying systemic disease. The features of pyoderma gangrenosum are not specific histopathologically and for this reason the diagnosis is based on clinical feature. The systemic administration of corticosteroids is the mainstay of treatment. We present five cases of pyoderma gangrenosum of age group ranging from 2 to 75 years. One of these patients was HIV-infected, and one was having discoid lupus erythematosus. The lesions were present on thighs in 3 cases, over back in one and breast in one. Biopsy showed central necrosis with multiple neutrophilic abscesses in epidermis. The dermis showed dense neutrophilic infiltration in both superficial and deep dermis. The lesions responded well to oral corticosteroids except in the case of HIV-infected patient in which minocycline showed considerable improvement.

Keywords: Minocycline, Prednisolone, Pyoderma gangrenosum

INTRODUCTION

Pyoderma gangrenosum is an uncommon ulcerative cutaneous condition with distinctive clinical characteristics and a frequent association with systemic disease. It was also known as phagedena geometrica, dermatitis gangrenosa, and phagedenic pyoderma. Pyoderma gangrenosum was first described and named by Brunsting et al. in 1930. They believed that streptococcal infection was a significant component leading to secondary cutaneous gangrene and named it as pyoderma gangrenosum. Hence, the term pyoderma gangrenosum is a misnomer. The exact causative mechanism is unknown. It is frequently associated with autoimmune diseases, but their exact role in producing the lesions is not known. The predisposed patient experiences an inciting event like minor trauma, but instead of a normal response that recognizes and removes the damaged tissue, the patient’s abnormal response results in the lesions of pyoderma gangrenosum.

The diagnosis mainly depends on recognition of the evolving clinical features because the histopathological changes are not specific. It is essential to exclude other diagnoses of cutaneous ulcers before therapy is initiated.

CASE REPORTS

Case 1
A 2-year-old female child presented with a huge ulcerative lesion over the right thigh with a history of insect bite 20 days back, after which a small, solid, reddish, round lesion appeared that ruptured, ulcerated, and acquired the present size within 10-12 days. The ulcer was painful and was gradually increasing in size. On examination, a single, well-circumscribed, oval-shaped ulcer measuring 8 cm × 5 cm in size, having violaceous borders, sloping edges, and indurated base was present over the anteromedial aspect of the right thigh. Floor was covered with yellowish slough with few blood-tinged areas (Figure 1).

Case 2
A 30-year-old male patient came with a history of trauma 12 months back over the left thigh which was followed by development of two ulcers with severe intractable pain, high-grade fever, and arthralgia. The ulcers were gradually increasing in size. On examination, two well-defined, irregularly shaped ulcers measuring 7 cm × 4 cm and 8 cm × 6 cm in size were present over the anterior aspect of left thigh and left side of lower abdomen, with presence of characteristic violaceous border. Ulcers were adjacent and separated by a band of normal skin. Floor was erythematous with few purulent areas in between. Edges were sloping and sharply defined (Figure 2).
Case 3
A 45-year-old HIV-positive male on anti-retroviral treatment, came with complaint of eruption of a single painful boil over the right thigh 1-month back, which ruptured leading to formation of a large ulcer which was gradually increasing in size. On examination, a single ulcer was present over the right iliac region and anterior aspect of right thigh measuring 8 cm × 4 cm with well-defined, sharp, blue to purple borders, sloping edges and erythematous floor with presence of granulation tissue (Figure 3).

Case 4
A 75-year-old male of discoid lupus erythematosus presented with a history of two pus-filled round lesions over the back after a surgical procedure. Within few days there appeared a small ulcer which gradually increased in size and acquired the present size. Mild pain was present over the site that had no aggravating factors. On examination, two sharply defined irregularly shaped ulcers were present over the back, measuring 5 cm × 3 cm and 6 cm × 4 cm in size with violaceous margins, erythematous floor, and sloping edge. Mild tenderness was present (Figure 4).

Case 5
An 18-year-old female patient presented with a history of nipple trauma 1-month back while feeding the baby. This resulted in a small wound which rapidly increased in size and led to the formation of a big ulcer. The ulcer was painful and was associated with serous discharge. On examination, a single well-defined oval ulcer of size 12 cm × 18 cm, with regular undermined edges and violaceous border was present over the right breast. Floor was covered with red granulation tissue, base was indurated and tenderness was present (Figure 5).

In all the cases, biopsy was done from the edge of the ulcer and was consistent with pyoderma gangrenosum, characterized by central necrosis with multiple neutrophilic abscesses in epidermis. The dermis showed dense neutrophilic infiltration in both superficial and deep dermis. Treatment was initiated with oral prednisolone at a dose of 1 mg/kg body weight in all the patients. Addition of oral

Figure 1: Pyoderma gangrenosum: A well-defined ulcer with a characteristic raised violaceous border in a 2-year-old female child

Figure 2: Pyoderma gangrenosum: Two ulcers measuring separated by a band of normal skin present over the anterior aspect of left thigh and lower abdomen

Figure 3: Pyoderma gangrenosum: A single ulcer over the right lower abdomen and thigh, with sloping edges and violaceous borders in an HIV-positive male

Figure 4: Pyoderma gangrenosum: Two well-defined ulcers with characteristic violaceous borders present over the back
minocycline at a dose of 100 mg twice daily was done in the HIV positive patient who was refractory to treatment. Standard wound care with daily cleaning and dressing of the ulcer was done followed by topical application of 2% gentian violet. Intravenous antibiotics were also given according to the culture sensitivity report. After 4-8 weeks of treatment, the wound size decreased, and healthy granulation tissue started appearing. Pain subsided, and the ulcers started healing.

**DISCUSSION**

Pyoderma gangrenosum is a primarily sterile inflammatory neutrophilic dermatosis. It is characterized by recurrent cutaneous ulcerations with mucopurulent exudate. These painful ulcers present with undermined bluish borders with surrounding erythema. Both humoral and cell-mediated abnormalities are associated with pyoderma gangrenosum. Hyperactivity of neutrophils and monocytes and hypersecretion of tumor necrosis factor - α has been found.

The peak of incidence occurs between the ages of 20 and 50 years with women being more often affected than men. Cases in infants and adolescents account for only 4%. Pyoderma gangrenosum in elderly people has occasionally been reported. The general incidence has been estimated to be between 3 and 10 per million per year.

Pyoderma gangrenosum occurs most commonly on the lower legs, mainly the pretibial area. It has been reported on other sites of the body as well, including breast, hand, trunk, head and neck, and peristomal skin. It is a feared complication after breast surgery and other surgical procedures. The ulcer starts as a follicular pustule with rapid growth, tissue necrosis, and enlargement of the area. The surrounding skin is erythematous, and the borders are typically undermined and violaceous or bluish. A strong sensation of pain is often associated with it. In at least 50% of patients, it is associated with systemic diseases such as inflammatory bowel disease, arthritis, monoclonal gammapathy, myeloproliferative disorders, acne conglobata, malignancies, and a host of other conditions like HIV, connective tissue disorders, sarcoidosis etc.

Clinically, pyoderma gangrenosum is classified into the following five varieties: Ulcerative type, pustular type, bullous type, peristomal type and vegetative type, with ulcerative type being most common. Diagnosis relies on clinical signs first and is supported by histopathology. Knowledge of the patient’s history for possible underlying disease and specific investigations based on that background are necessary. Therefore, the diagnosis is made by exclusion of other possible disorders. No laboratory parameter for pyoderma gangrenosum is available. Histopathology is non-specific. The initial lesions show a deep suppurative folliculitis with dense neutrophilic infiltrate. In about 40% of cases, leukocytoclastic vasculitis is present. Pyoderma gangrenosum with (necrotizing) granulomatous inflammation has been described.

Treatment is directed toward underlying associated systemic disease, immune system contributions, and the local wound. Steroids given orally or intravenously are effective in treating systemic immune contributions. Refractory cases have been successfully treated with cyclosporine, clofazimine, dapsone, thalidomide, azathioprine, and other immunosuppressive agents. Combination therapies of systemic steroids and hyperbaric oxygen in the treatment of wounds associated with pyoderma gangrenosum are also successfully reported.

**CONCLUSION**

Diagnosis of pyoderma gangrenosum is mainly clinical, and the mainstay of therapy is systemic corticosteroids but minocycline can also give good results. These cases are presented for their rarity, a wide spectrum of presentation (2-75 years) and the association with systemic conditions.

**REFERENCES**


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