

Pyoderma Gangrenosum of Abdominal Wall

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P yoderma gangrenosum is an ulcerative disease of skin, the etiology of which is obscure. The authors are reporting a case of pyoderma gangrenosum of the abdominal wall that was initially misdiagnosed as a parietal wall abscess and exhibited characteristic pathergy after drainage. Diagnosis was made on clinical grounds and by the presence of pathergy. Abdominal wall pyoderma gangrenosum should be suspected in any patient with non-healing ulcerative lesions of the abdominal wall. The patient responded well to steroids with no recurrence seen in the follow up period.

Pyoderma gangrenosum, a neutrophilic dermatosis, is a rare ulcerative cutaneous disease. An immunological abnormality has been proposed as a possible mechanism. There are no specific clinical features seen with this disease. This has characteristic features of pathergy. Diagnosis relies on excluding other causes of ulcerative skin diseases. The occurrence of pyoderma gangrenosum at a surgical site is rare especially if there is no predisposing illness. Early diagnosis is essential to prevent pathergy. Corticosteroids are the main drugs used for treatment.

A 28 year old female presented with painful swelling of the right abdominal wall and fever of 5 days duration for which she had received antibiotics but had no relief, with the worsening of symptoms and progressive increase of the swelling. On general physical examination, the patient had a dehydrated look and fever of 100 degrees Fahrenheit. Systemic examination was normal. Local examination revealed a tender swelling measuring 10 × 7.8 × 2.3 centimeter showing fluctuation, free from underlying structures, with prominence on Carnett's test's (leg raising test) in the right lower abdomen and was diagnosed as right parietal wall abscess, and had an incision and drainage. Thirty-six hours after incision and drainage, and despite improvement, there was an ulcer with pus and development of another pustule, and so the patient underwent another debridement. There was seen further worsening of symptoms despite the patient being on broad spectrum antibiotics, and the drainage of abscess were no improvement and the patient developed a third ulcerated lesion adjacent to the previous ones. The ulcers had a dirty floor with asymmetrical borders (Fig.1). Histopathological examination identified a non-specific mixed inflammatory cell infiltrate with

areas of necrosis. Cultures of the debrided tissue were negative for bacteria, mycobacteria, and fungi. Serum protein electrophoresis was normal, antiphospholipid, antinuclear and antineutrophil cytoplasmic antibodies and rheumatoid factor was negative. No associated disease was identified. The patient was diagnosed as a case of Pyoderma gangrenosum and so was put on corticosteroids. He had a good response and had the lesion healed. No fresh recurrence was seen after follow up of 9 months.



Figure 1: Ulcers having dirty bases with asymmetrical borders (Pyoderma Gangrenosum)

Pyoderma gangrenosum was first reported in 1924 following drainage of an abdominal abscess and is regarded as a diagnosis of exclusion.¹ It is categorized into four types; ulcerated pyoderma, bullous, pustular and vegetans, each with its own peculiarities and different clinical responses to treatment. The ulcerative form of pyoderma gangrenosum is characterized by a rapidly progressing painful irregular and undermined bordered necrotic ulcer.²

Lesions can be solitary or multiple, chronic or recurrent. Any site on the body is prone to developing this skin dermatosis. They commonly occur on the legs, especially on the pretibial area, face, neck, scrotum, or penis. Pyoderma gangrenosum of the abdominal wall adjacent to stoma following colectomy can occur. Post-surgical pyoderma gangrenosum can affect any surgical wound. However, the abdominal wall and the breasts are the most affected areas.³

Approximately 50% of the cases of Pyoderma gangrenosum occur concomitantly with other diseases, notably with systemic diseases especially arthritides, inflammatory bowel disease and hematological disorders (leukemia) disorder, with inflammatory

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bowel disease being the most common association. Less commonly associated diseases include hepatic diseases, myelomas, immunological diseases and HIV infection.⁴ The remaining forms occur in an isolated manner. Lesions can develop spontaneously, after surgery or trauma. An immunological anomaly of the hyperergic reaction type, involving IL-8 (interleukin-8) and IL-16 (interleukin-16) have been implicated in etiology of Pyoderma gangrenosum. Mixed genetic predisposition, undefined infectious agents or paraneoplastic or paraimmune phenomenon are the factors maintaining these abnormalities.⁵

Pyoderma ulcers are classically painful and aseptic, although super infection may eventually occur. The sequence of events in the formation of this ulcerative lesion starts initially with a pustule or inflammatory nodule, which breaks down to form an ulcer. This ulcer shows purulent base with ragged undermined asymmetrical border with sometimes a purple-red edge which, spreads centrifugally. The distinctive clinical features of pyoderma gangrenosum are apparent enough to permit the diagnosis of most cases, comprising in its classic form a burrowing ulcer with an irregular margin and ragged purple-red overhanging edge. The extension of lesions of Pyoderma gangrenosum in response to trauma or surgical debridement, termed pathergy, is a hallmark of this entity, and is observed in 25–50% of cases.⁶ Sometimes multiple ulcers of pyoderma gangrenosum may coalesce resulting in a large ulcer, which usually heals with a pigmented cribriform scar. Histopathological examination of the ulcer varies with the stage of the disease. In early cases, neutrophilic abscess is seen followed by epidermal necrosis and ulceration, with a superficial dermal edema and a dense mixed dermal infiltration in later stages whereas biopsy of advancing inflamed border demonstrates perivascular lymphatic inflammation.

The rarity of the disease, the condition's ability to mimic other ulcerative lesions and its lack of specific laboratory and pathologic findings leads to misdiagnosis and mistreatment of the lesion. A high index of clinical suspicion can lead to a definitive diagnosis. The frequency of misdiagnosis is about 10%. Its treatment is still a motive for discussion in the literature with no uniformly effective or specific therapy. Immunosuppression is the basis of treatment. Debridement or necrosectomy in postoperative Pyoderma gangrenosum is contraindicated.⁷

Corticosteroids and cyclosporine are the most commonly used drugs as immunosuppressants. Topical treatment includes topical or intralesional corticosteroids, tacrolimus ointment, intralesional cyclosporine, topical 5-aminosalicylic acid, nitrogen mustard or 0,5% nicotine cream.⁸ These topical agents are generally used as supportive treatment for the systemic treatment and their use as monotherapy has a limited role. In cases without associated systemic disease, intravenous immunoglobulins, granulocyte and monocyte adsorption apheresis, plasmapheresis and cyclophosphamide treatment have been reported to be effective.

Pyoderma gangrenosum of the abdominal wall is a rare disease. It can be easily misdiagnosed with other ulcerative skin diseases and pathergy is important for diagnosis.

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