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Pustular Pyoderma Gangrenosum with Ulcerative Colitis an Uncommonly Seen Association.

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Abstract

We report a case of a 36-year-old female having ulcerative colitis who had multiple papulo-pustular lesions over trunk and face. She had received treatment as acne vulgaris but due to lack of satisfactory response, histopathological evaluation was done which was consistent with pustular pyoderma gangrenosum. The diagnosis of pustular pyoderma gangrenosum is often challenging as there is no defining diagnostic clinical, laboratory or histopathological feature. Thus a high index of suspicion is essential to diagnose this condition.

Key words: Papules; Pustular; Pyoderma gangrenosum; Ulcerative colitis.

Introduction

Pyoderma gangrenosum (PG) is an idiopathic inflammatory disease of unknown etiology, commonly associated with an underlying systemic condition such as hematological malignancy or inflammatory bowel disease. Its incidence tends to parallel exacerbations of the underlying disease. Four clinical variants of PG have been described, and these include ulcerative, pustular, bullous, and vegetative types, of which ulcerative pyoderma gangrenosum is the most common type. [1] We report a case of a 36 year old female patient with ulcerative colitis associated with pustular pyoderma gangrenosum.

Case report

A 36-year-old female presented with multiple discrete erythematous, painful papules that rapidly developed into pustules on her face, back, upper chest, arms, and thighs for the past 5-6 years. These pustules subsided by themselves in 1-2 weeks leaving behind postinflammatory hyperpigmentation without any ulcers. New lesions kept on developing on and off with no clear aggravating factor. Since the past 1 year her papulopustular lesions have increased in number. She sought treatment from multiple doctors, who diagnosed her with acne vulgaris. She was treated for the same with oral doxycycline, topical clindamycin solution, benzoyl peroxide gel 5%,topical ketoconazole shampoo, but it did not yield satisfactory results,and new lesions continued to develop. The patient visited our hospital seeking treatment for the same complaints. The skin lesions have aggravated for the last 1 year and there is a history of multiple episodes (every 3 to 4 months) of abdominal pain associated with diarrhea and weakness since the last year but these gastrointestinal symptoms are not always associated with the aggravation of skin lesions. There is no history of fever, malaise, arthralgia, or burning on skin lesions. On examination, she was found to have multiple erythematous papules and pustules over her face, back, upper chest, arms, and thighs. Many small hyperpigmented macules were also present which were suggestive of post inflammatory hyperpigmentation

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and scars (Figures 1 & 2). The patient was a known case of ulcerative colitis, which was diagnosed in 2015 based on clinical symptoms, a colonoscopy, and biopsy findings. A colonic biopsy showed distortion of mucosal architecture with inflammatory infiltrate consisting of neutrophils, lymphocytes, eosinophils, and plasma cells in the lamina propria. Cryptitis and crypt abscesses were also identified. She had been receiving treatment from a gastroenterologist but had stopped all medications for the past 1 year. Provisional diagnoses of cutaneous manifestations associated with ulcerative colitis include most probably pyoderma gangrenosum, pustular Sweet's syndrome, and sub corneal pustular dermatosis. These diagnoses were made on the basis of clinical history and underlying systemic disease. Oral and skin pathergy tests were done using 18G hypodermic disposable needle inserted in the dermis at the flexor aspect of the left forearm and lower lip mucosa, respectively and read after 48 hours, which came out negative. All hematological investigations, including renal and liver function tests, were normal except for anemia. The swab for pus culture from the pustule did not show any growth. A 4 mm punch skin biopsy was done on a pustular lesion on her lower back. Histopathology showed normal epidermis with dense subepidermal perivascular neutrophilic and lymphoplasmacytic infiltration along with dermal collagenosis, which was consistent with pyoderma gangrenosum (Figure 3). The patient was then prescribed tablet prednisolone 25mg for 20 days and tapered to 5 mg over a period of 3 months, along with oral mesalamine 1.2 gm and tablet rifaximin 400 mg for ulcerative colitis according to opinion of gastroenterologist, which resulted in dramatic improvement in skin lesions. There was improvement in the lesions within 5-10 days after starting oral prednisolone and no new papulo-pustular lesions developed during the 3 - months follow up of the patient. The patient did not have any active gastrointestinal complaints for her ulcerative colitis.

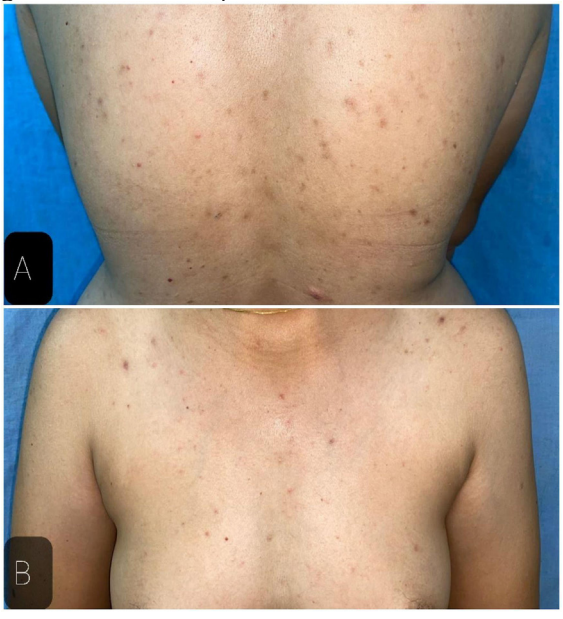

Figure1: Multiple discrete erythematous papules with post inflammatory hyperpigmentation and scars over back (A) and chest (B) seen.

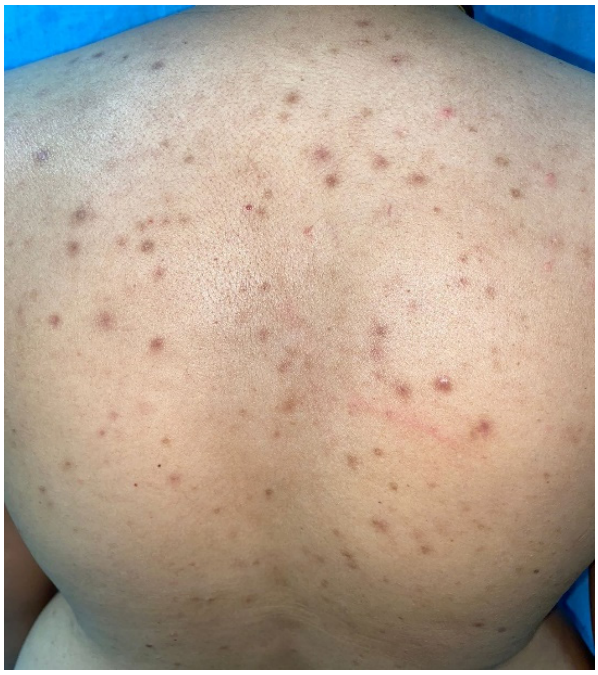

Figure2: Multiple discrete erythematous papules with post inflammatory hyperpigmentation and scars over back seen.

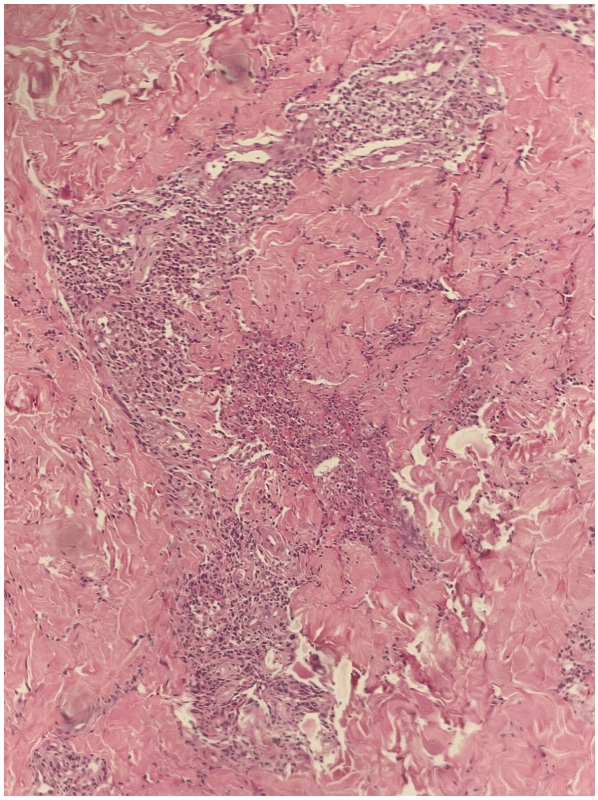

Figure3: H & E stained smear in 40× magnification from lesion over back showing (black arrow) polymorphonuclear cell infiltration, lymphocyte , plasma cell , histiocytes in dermis with subtle necrosis. Dermal collagenosis (blue arrow) is also seen.

Discussion

Each variant of pyoderma gangrenosum (PG) exhibits distinct clinical and histopathological features associated with different conditions (Table 1); hence prognosis and treatment are different for each variant. While typically only one specific type of pyoderma gangrenosum (PG) occurs in an individual, there are instances where combinations of different clinical types may arise. The pustular form of PG was initially identified by O'Loughlin and Perry in two patients with acute ulcerative colitis. [2] These patients developed a widespread pustular eruption that did not progress to ulceration. The pustules were described as painful and mainly located on the extensor extremities and upper trunk, occurring during severe exacerbations of colitis and accompanied by fever and arthralgia. Categorizing PG based on its predominant morphological features is beneficial in determining the likelihood and type of underlying systemic disease (Table 1). [3] Pustular PG is frequently seen in individuals with ulcerative colitis, occurring at a rate of 1-6 %. The course of pustular PG often mirrors the severity of bowel symptoms, manifesting primarily during colitis exacerbations. It can serve as a marker for bowel disease activity and tends to improve with treatment of the underlying inflammatory bowel disease. [4] However, PG can also manifest itself when colitis symptoms are absent or minimal, as was seen in our patient. [5] The diagnosis of PG is primarily clinical, as there are no specific histopathological or immunofluorescence patterns that define the condition. Making the diagnosis can be challenging and relies on recognizing the broad spectrum of clinical features, identifying systemic disease associations. Differential diagnoses that should be kept in mind include acne vulgaris, pustular sweet's syndrome, and sub corneal dermatosis. A lack of response to standard anti acne treatment, involvement of extremities, absence of fever, lack of tissue, or peripheral neutrophilia help rule out these diagnoses. Although the histopathological features are not pathognomonic, a skin biopsy is necessary to exclude other causes of acute skin ulcerations, particularly infections and necrotizing fasciitis. In pustular PG, characteristic findings include subcorneal neutrophilic accumulation, [6] perifollicular neutrophilic infiltration, and subepidermal edema with dense dermal neutrophilic infiltrate. These changes reflect various stages of the disease, likely influenced by the timing of the biopsy relative to the disease evolution. A stepwise approach to treating pyoderma gangrenosum involves first controlling lesions with high-dose glucocorticoids; typically, four to six weeks of glucocorticoid therapy are required, followed by tapering off glucocorticoid-sparing agents. Cyclosporine can be an effective alternative to prednisone and has been used successfully in combination with mycophenolate mofetil. Agents that have been used as glucocorticoid-sparing agents include dapsone, minocycline, azathioprine, tacrolimus, mycophenolate mofetil, methotrexate, chlorambucil, and clofazimine. [7] Tumor necrosis factor-alpha inhibitors, such as adalimumab or infliximab, have also been shown to be helpful in refractory disease, especially in patients with inflammatory bowel disease. Intralesional glucocorticoids and topical tacrolimus have also been used in treatment. [7]

Table 1. Clinical and histopathological features of PG Variants [3.]

	Clinical features	Histopathological features	Associated Systemic Disease
Ulcerative	Rapid progression of a painful ,necrolytic cutaneous ulcer with an irregular ,violaceous , and undermined border History suggestive of pathergy or clinical finding of cribriform scarring Systemic disease associated with PG Treatment response(rapid response to systemic steroids)	Sterile dermal neutrophilia, mixed inflammation,lymphocytic vasculitis.	Inflammatory bowel disease, Rheumatoid arthritis, Monoclonal gammopathy, Malignancy particularly myeloproliferative disorder eg.leukemia,myelodysplasia,polycythemia rubra vera.
Pustular	Painful pustules(0.5-2 cm diameter), with surrounding halo Associated inflammatory bowel disease Improvement with control of inflammatory bowel disease Systemic steroid are the mainstay of treatment	Neutrophilic infiltrate; Subcorneal /subepidermal neutrophils.	Very frequent: inflammatory bowel disease(ulcerative colitis) Rarely: Polycythemia rubra vera, Hepatobiliary disease, pyostomatitis vegetans.
Bullous	Painful ,inflammatory bullae;rapidly enlarging painful vesicles and bullae;coalescing of grouped bullae Associated hematologic malignancy in as many as 70% Pathergy Rapid response to steroid	Neutrophilic dermal infiltrate, subepidermal bullae,epidermal necrosis.	Frequently present: Myeloproliferative disorders.

Vegetative	Chronic erythematous plaques ,with sinus formation ;shallow ulceration or erosion and discomfort No associated disease Response to minor treatment measure(topical and intralesional steroid and less commonly ,oral corticosteroid and other immunosuppressive agent)	Dermal and histiocytic dermal infiltrate, granuloma formation.	Usually no associated disease .

Conclusion

Due to its rare occurrence, pustular pyoderma gangrenosum (PG) may often be misdiagnosed with other diseases. Vigilance during history taking is necessary to identify any underlying systemic condition. Additionally, alternative diagnoses should be considered in patients who present with non-responding papulopustular lesions in the background of inflammatory bowel disease.

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