PYODERMA GANGRENOSUM IN A CHILD: A RARE CASE

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ABSTRACT

Pyoderma gangrenosum, a rare neutrophilic dermatoses commonly occurs between 3rd to 5th decades of life. We report a case of pyoderma gangrenosum in a child of 9 years who presented with a non-healing ulcer refractory to conventional therapies with positive pathergy test. Histopathologic examination revealed marked diffuse dermal neutrophilic infiltrate. Rapid improvement with Tab. prednisolone 1 mg/kg/day was observed within 15 days.

KEYWORDS
Pyoderma Gangrenosum, Child, Pathergy Test

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatoses, first described by Brocq in 1916, which is non-infectious and commonly associated with an underlying systemic disease.1 It is characterized by sterile neutrophilic infiltration of the skin and other organs.2 Most patients develop PG between 25 and 45 years of age.3 We report a case of non-healing ulcer refractory to conventional therapies in a 9 year old child which later diagnosed as PG with excellent response to oral corticosteroids.

CASE REPORT

A 9 years old male child presented with a solitary painful ulcer over the posterior aspect of right thigh which had evolved from a small tender red swelling over 3 months. Pain was out of proportion to the size of ulcer and impaired walking and other day to day activities. It was gradually progressing. There was no improvement despite treatment with antibiotics for prolonged period.

On general examination, patient appeared normal except for moderate pallor & poor nutrition. There was no lymphadenopathy or swelling of limbs.

On cutaneous examination, the ulcer was ovoid in shape around 15 cm in diameter extending from mid-thigh above to popliteal fossa below; base was filled with granulation tissue and necrotic material and edge was sharply demarcated, violaceous and undermined. (Figure 1) Palpating the ulcer revealed exquisite tenderness. Further examination revealed, there was a 1 cm sized papule on the dorsum of right hand at the site of intravenous cannulation suggestive of positive pathergy test. (Figure 2).

Fig. 1, 2: Ulcer over the posterior aspect of the right thigh with violaceous and undermined edge- Initial presentation. Figure 2 also showing positive pathergy phenomenon at the intravenous cannulation site over dorsum of right hand

Cutaneous examination of rest of body and systemic examination did not reveal any abnormalities.

Routine laboratory tests of blood, urine and stool were normal apart from Haemoglobin was 9.6 gm%; total leukocyte count (TLC) 10700 cells/cubic mm; erythrocyte sedimentation rate (ESR) 40 mm at the end of first hour; Tridot screening for Human Immunodeficiency Virus (HIV) was negative. Sputum for AFB and Mantoux test was negative, TB interferon was also negative. Chest X-ray and other imaging was within normal limits.

Biopsy was taken from edge of ulcer. Histopathological examination showed marked diffuse dermal neutrophilic infiltrate with a variable lesser infiltrate of lymphocytes and histiocytes with epidermal necrosis. (Figure 3)

Fig. 3: Histopathology showed marked diffuse dermal neutrophilic infiltrate with a variable lesser infiltrate of lymphocytes and histiocytes with epidermal necrosis. (H & E ×10)

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A final diagnosis of Pyoderma gangrenosum was made and the patient was put on Tab. prednisolone 1mg/kg/day. Within 5 days of starting treatment patient improved symptomatically. Ulcer started healing rapidly and within 15 days of starting treatment ulcer healed with reduction of size. (Figure 4)

DISCUSSION
PG is an inflammatory neutrophilic dermatosis characterized by ulceration of skin often associated with underlying systemic disorder. M: F ratio is 1:1. PG developed in most of cases between 25-45 years and cases in children are unusual.

Exact aetiology is unknown. Multiple abnormalities of the cellular and humoral immune responses along with neutrophil abnormalities have been described, but the pathogenic role of these changes remains uncertain. However it is seen in association with Inflammatory bowel disease e.g. ulcerative colitis and Crohn’s disease, joint disorder e.g. rheumatoid arthritis, seronegative arthritis and osteoarthritis, haematological disorders (Leukaemias, myelofibrosis, myelodysplastic syndromes, polycythaemia vera, paraproteinaemia, monoclonal gammopathy, Waldenström’s macroglobulinaemia, paroxysmal nocturnal haemoglobinuria), connective tissue disorders, vasculitides, infections, drugs like G-CSF etc.

PG may present with four clinical variants 1) classic or ulcerative type 2) Pustular 3) bullous and 4) vegetative type. Classic or ulcerative type is the commonest type starts as a painful nodule or folliculo athergy). The indurated border may be seen as studded with neutrophil abnormalities have been described, but the pathogenic role of these changes remains uncertain. When associated with Crohn’s disease granuloma with giant cells may be seen.

Diagnosis is based on clinical features and course along with characteristic histological features. The differential diagnosis should include gangrene, mycobacterial infections, ecthyma gangrenosum, deep mycoses, tropical ulcer, factitial ulcers, halogenoderma, pemphigus vegetans, spider bites, stasis ulcer, and Wegener’s granulomatosis. Appropriate biological testings are very important to exclude other differentials along with clinico-pathological correlation.

The first and foremost step in the management of PG is to search for any underlying systemic disease and treat it accordingly which is not only necessary for maintaining remission but also prevent relapse.

For limited and mild disease, local therapy with topical/intralesional steroids or topical tacrolimus may be sufficient. Systemic corticosteroids are the mainstay of the therapy for the acute, rapidly progressive form of this disease. Initial doses of prednisolone in the range of 40–80 mg/day or higher are usually required to control acute flare. In some cases pulse steroid therapy may produce rapid improvement of disease that was unresponsive to oral corticosteroids.

Other immunosuppressive agents like azathioprine, cyclophosphamide, chlorambucil, Mycophenolate mofetil and colchicine can be used. Sulfasalazine (1–4 g/day), sulfapyridine, sulfamethoxypyridazine, dapsone (100–200 mg/day) 1,2,3 have been successfully used in the management of PG. Biologics like Infliximab also has given dramatic results in the healing of lesions.

REFERENCES