IDIOPATHIC PYODERMA GANGRENOSUM: REPORT IN A THREE YEAR OLD CHILD

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HOW TO CITE THIS ARTICLE:
Vidyadhar A. Kinhal, Sanjeev B. Joshi, Sambasiviah Chidambara Murthy, Syeda Siddiqua Banu. "Idiopathic Pyoderma Gangrenosum: Report in a Three Year Old Child". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 3, January 19, 2015; Page: 272-277.

ABSTRACT: Pyoderma gangrenosum is a rare, non-infectious and destructive skin disease. Its exact etiopathogenesis is not clearly understood and is believed to be due to immune dysfunction. Several theories have been postulated; however none are consistent in all patients. Pyoderma gangrenosum mostly affects adults and rarely in children. An underlying systemic disease is often seen in most of the cases. A three- year- old male child presented with a non-healing ulcer over left thigh. Clinical features, histopathology, other investigations and dramatic response to corticosteroids suggested a diagnosis of idiopathic pyoderma gangrenosum. Associated idiopathic thrombocytosis was also seen. Idiopathic pyoderma gangrenosum is very rare in children. Idiopathic pyoderma gangrenosum associated with idiopathic thrombocytosis and keloidal scarring has not been reported earlier.

KEYWORDS: pyoderma gangrenosum, idiopathic, skin ulcers, children, corticosteroids, immuno suppressive agents.

INTRODUCTION: Pyoderma gangrenosum (PG) is an uncommon, non-infectious destructive skin disease. The peak incidence occurs between the ages of 20-50 years with a slight female preponderance.[1,2] An incidence of only 4% is seen in children.[3]

The precise etiopathogenesis is not well understood; however immunological factors and neutrophil dysfunction are thought to have an important role. More than 50% of patients have an associated systemic disease, which may precede, follow or occur simultaneously [4] Idiopathic variety is rare and diagnosis should be made after ruling out the presence of all systemic associations. We report this rare entity in a three year old male child associated with idiopathic thrombocytosis and keloidal scarring.

CASE REPORT: A three- year- old male child presented to the emergency ward with a painful ulcer over his left thigh (Figure. 1) of one week duration that was preceeded by a small pustule and mild fever. There was no history of trauma, significant past medical or surgical history. On general physical examination child was anxious and febrile; caput quadratum, slight bowing of legs was noted, pallor, edema, icterus, nail clubbing and lymphadenopathy were absent and systemic examination was normal. Child was put on intravenous antibiotics; local wound care with surface antiseptic and daily dressings were done, calcium supplements were added for nutritional rickets after a pediatric opinion.

Despite intravenous antibiotics and daily dressings for one week, the child showed no signs of improvement. The ulcer was spreading rapidly approximately at the speed of 1 cm per day encircling more than half of the thigh circumference and spread below the left knee

(Figure. 2). Subsequently, he also developed similar lesions over gluteal region (Figure. 3), chest, right upper limb, that started as a pustule and progressed into ulcers; pathergy phenomenon was noted as the lesions also appeared at the sites of venupuncture (Figure. 4). The ulcers were similar; except for the variation in size. All ulcers had necrotic base, irregular and raised inflammatory edges, minimal serous discharge without lymphadenopathy.

Investigations Revealed: mild anemia (Hb: 8.4gm), with packed cell volume: 30%, total leucocyte count: 22, 000 cells/cm^3. erythrocyte sedimentation rate: 50 mm at the end of one hour and platelet count 10, 63, 000/cm^3. Serum calcium levels were low (total 7mg/dl, ionized 4mg/dl) fasting, post prandial blood sugar levels and thyroid profile was normal. The peripheral smear showed microcytic hypochromic anemia, neutrophilic leucocytosis and thrombocytosis, with no abnormal cells. Abdomino-pelvic ultrasonogram and chest radiograph were normal. Antinuclear antibody, enzyme-linked immunosorbent assay for Human Immuno deficiency Virus 1 and 2, hepatitis C virus, hepatitis B surface antigen and blood culture and stool examination were all negative. Barium enema was normal.

Gram's staining, tzanck smear, fungal cultures from the edge of the ulcer were negative, bacterial swabs from the edge of the ulcers showed "presence of E.coli "sensitive to Amikacin. Skin biopsy revealed dense mixed cell infiltration of neutrophils with areas of haemorrhage and necrosis without vasculitis (Figure. 5). Bone marrow biopsy showed non-specific megakaryocytic hyperplasia. A diagnosis of classic (Ulcerative) form of pyoderma gangrenosum was made, and child was started on high dose of intravenous methyl prednisone (15mg/day), and injection Amikacin as bacterial culture from ulcers grew E.coli sensitive to this antibiotic, calcium supplements were continued. Blood transfusion was done for the correction of anemia. Immediate response to corticosteroids was seen and lesions started healing (Figure. 6), subsequent bacterial swab cultures showed no growth and blood parameters normalised.

Corticosteroids were continued and within ten days all lesions healed completely with keloidal scarring (Figure. 7). Child was discharged with oral corticosteroids and is under regular follow up.

DISCUSSION: The salient feature of pyoderma gangrenosum is an ulcer with a raised inflammatory border and a boggy, necrotic base predominantly affecting lower extremities in both adults and children. In children, preferential involvement of buttocks, perianal and genital region is seen along with involvement of head. **[3]** The mode of presentation of PG is variable. It might be either sudden in onset with rapid progression and severe necrosis or gradual progression with spontaneous regression.[5] Mucous membranes are usually spared but aphthous lesions may occur in the oral cavity. There is no lymphadenopathy or lymphangitis. Twenty percent of cases show Koebner phenomenon (or isomorphic response or pathergy) i.e: the development of a skin lesion similar or identical to the index lesion at the site of mechanical trauma.[5]

Ulcerative (Classic form) is the most common and well recognised form characterized by painful, large ulcerations with purulent base and undermined borders that require aggressive systemic immunosuppressive therapy for control. Other forms described are: Pustular, Bullous, superficial(vegetative), peristomal and Extracutaneous form involving Lungs, heart, central nervous system or other internal organs in the absence of cutaneous lesions.

The histopathological features depend on the type of the lesion, the stage of evolution and the site from which the biopsy specimen is obtained. Central necrosis and ulceration of the epidermis and dermis surrounded by an intense massive neutrophilic infiltration ("sea of neutrophils"), in the absence of vasculitis and granuloma formation is typical of PG.[6]

Prognosis depends upon the type as well as severity of associated disorders.[7] The commonest associated disease is arthritis in adults, and ulcerative colitis in children.[3] Other associated systemic diseases are:

1. Para inflammatory (paraimmune) (associated with inflammatory bowel disease, collagen vascular diseases, arthritis, autoimmune neutropenia in infancy, immunosuppression, thyroid disease, diabetes mellitus etc)
2. Paraneoplastic (associated with malignancy: colon, pancreas, breast, bronchus, carcinoid)
3. Hematologic (leukemias, polycythemia)
4. Drug induced (interferon, propylthiouracil, isotretinoin, etc)
5. Infectious (HIV, Hepatitis C).

However no underlying disease associations can be found in about 25%-50% of PG.[5-7]

The diagnosis of pyoderma gangrenosum should be made on the basis of the following diagnostic criteria:[2] both major and at least two minor criteria's should be present.

Major criteria are rapid progression of painful ulcers (usually >1 cm/day) with irregular; undermined edges and necrotic base and exclusion of other causes of ulceration. Minor criteria include pathergy phenomenon, presence of a disease with known association with pyoderma gangrenosum**,** appropriate histopathological findings, rapid response to oral corticosteroid therapy (usually interpreted as at least 50% reduction in size using 1–2 mg/kg/day).In our patient both the major and three minor criteria were present, based on which the diagnosis was made.

Corticosteroids and immunosuppressives are the most predictable and effective treatment modality against pyoderma gangrenosum. The treatment of underlying disease may aid in healing. Local therapy with wet compresses, saline and alginate dressings are an important adjunct to systemic therapy and may provide relief from symptoms **[8]**. Aggressive surgical debridement or skin grafting should not be routinely done because of the risk of a pathergic response.

The long-term outcome for patients with pyoderma gangrenosum remains unpredictable. So patients should be followed up for a longer period of time and corticosteroids should be continued for 3-6 months.

CONCLUSION: In conclusion, we report a rare occurrence of idiopathic pyoderma gangrenosum with idiopathic thrombocytosis and keloidal scarring in a very young child. High index of suspicion and knowledge of this destructive skin disease helps in timely diagnosis and intervention thereby preventing the associated morbidity and mortality.

ILLUSTRATIONS:

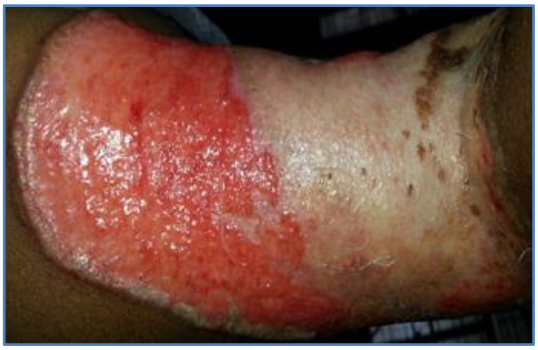

Fig. 1: Ulcer with irregular edges over left thigh on presentation

Fig. 2: Ulcer involving entire left thigh and knee

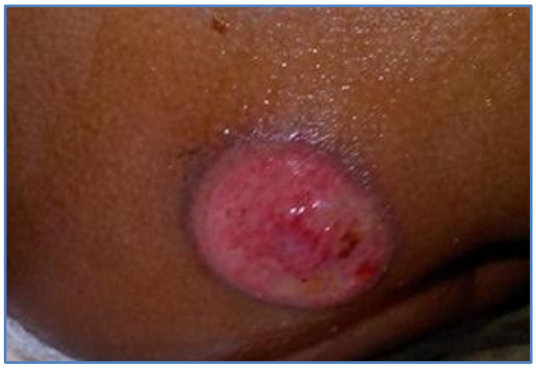

Fig. 3: Similar lesion over the gluteal region

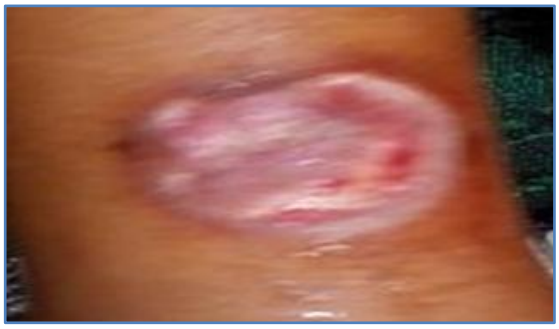

Figure 4: Pathergy over the IV canula sites

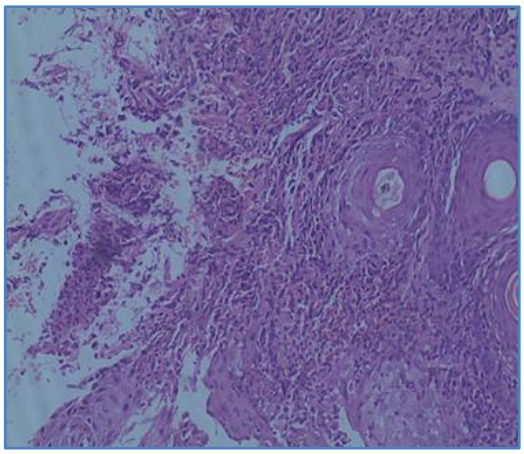

Fig. 5: HPE showing ulceration of the epidermis with dense dermal neutrophilic. infiltration without vasculitis and granuloma formation (H &E, x100)

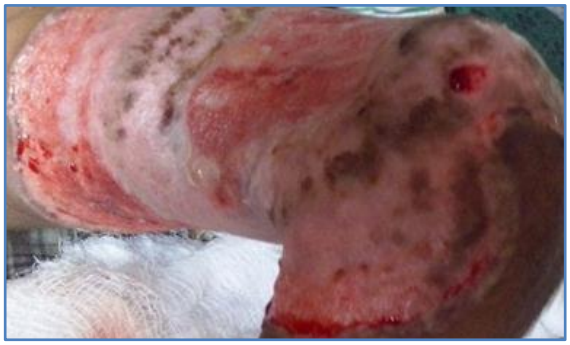

Fig. 6: Ulcer after starting systemic steroids

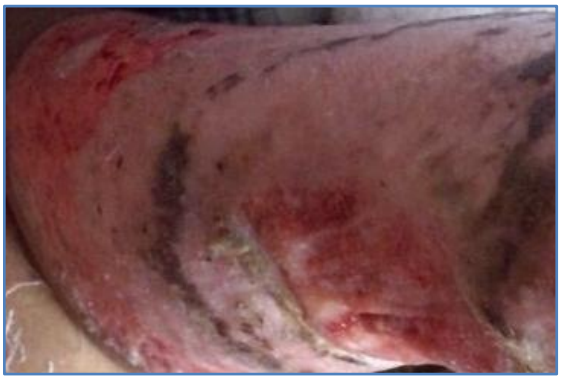

Fig. 7: Healed ulcer with keloidal scarring after 10 days of systemic corticosteroid therapy

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Date of Submission: 08/01/2015.
Date of Peer Review: 09/01/2015.
Date of Acceptance: 10/01/2015.
Date of Publishing: 17/01/2015.