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Pyoderma Gangrenosum in a Child - A Rare Case Report

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Abstract

Pyodermagangrenosum (PG) is a rare, reactive, chronic, non infectiousneutrophilicdermatosis of unknown etiology commonly associated with underlying systemic diseases. It is seen more frequently in adults between 40-60 years, with female preponderance and affects only 4% of infants and adolescents. Here we report a case of pyodermagangrenosum in a child which is relatively rare.

Keywords: Pyodermagangrenosum, child, perianal, perineum, pathergy

Introduction

Brocq in 1916 first described the disease as "Phagedeniomegeometrique". Brunsting, Goeckerman, O' Leary in 1930 named it as "Pyodermagangrenosum" [1].

PG has many variants, classical ulcerative being the commonest. Many theories on its etiology has been postulated. Diagnosis of pyodermagangrenosum is based on specific criteria – Sue et al criteria which has 2 major and 4 minor criteria of which atleast 2 major criteria and 2/4 minor criteria should be present[2]. Exclusion of other causes of ulceration is a must. Histopathological findings are not diagnostic but essential to rule out alternative diagnosis.

Case Report

A 3 ½ year old, 2nd born, nonconsanginous, female child presented with multiple nonhealing ulcers over face and body for 2 ½ years duration. It started as itchy raised skin lesion which progressed to form ulcer over forehead slowly involving the lower limb, perineum, perianal area and upper limb.

- On general examination – child was thin, malnourished, awake and irritable, afebrile, pallor and pedal edema was present. Vitals were BP:110/80 mmHg, PR- 90/min.
- Systemic examination was normal.
- Dermatological examination revealed multiple ulcers of varying sizes 4x4cm to 15 x 8cm present over forehead (Figure 1), bilateral cubital fossa, thighs and legs; perineum (Figure 2) and perianal area(Figure 3). Ulcers showed violaceous and irregular border, undermined edge, floor covered with slough and granulation tissue, indurated base. Hair and scalp, oral mucosa, nails were all normal. Pathergy test was found positive at the site of intravenous cannula.

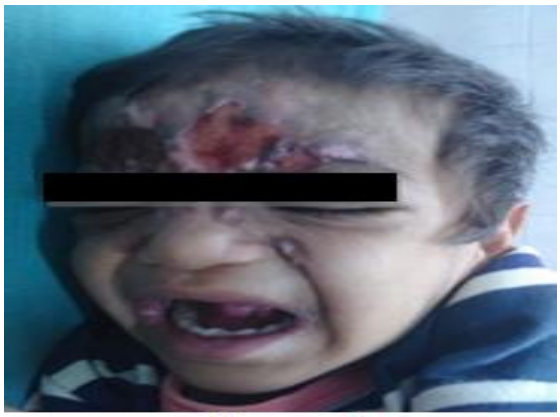

Figure 1

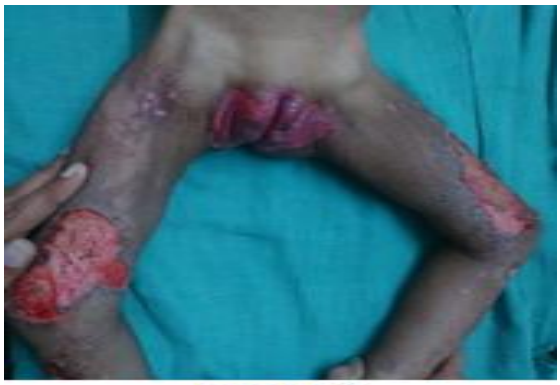

Figure 2

Figure 3

Gastroenterology and hematology opinion was sought to rule out associated inflammatory bowel disease and hematological malignancies and no evidence involving respective system was found.

A provisional diagnosis of pyodermagangrenosum was made.

Investigations

- Complete hemogram - anemia, neutrophilic leukocytosis, peripheral smear - nisopoikilocytosis, LFT- hypoalbuminemia, RFT- hypokalemia,

- Other tests - ANA, immunoglobulin levels, viral markers,VCTC, VDRL, USG abdomen, stool examination, sickling test, spot protein creatinine ratio, skeletal survey, urine RE, chest Xray, blood and urine culture &sensitivity - all were within normal limits.

- Histopathological examination from the edge of ulcer (right leg):

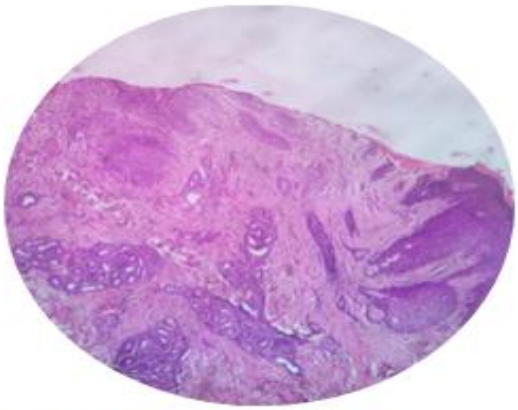

Figure 4- Ulceration of epidermis in one part and epidermal hyperplasia with spongiosis in another part (H&E, 10x)

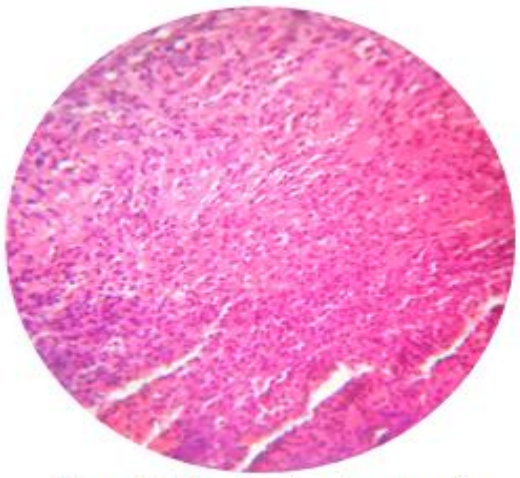

Figure 5 - Dermis showing sea of neutrophils (H& E, 40 x)

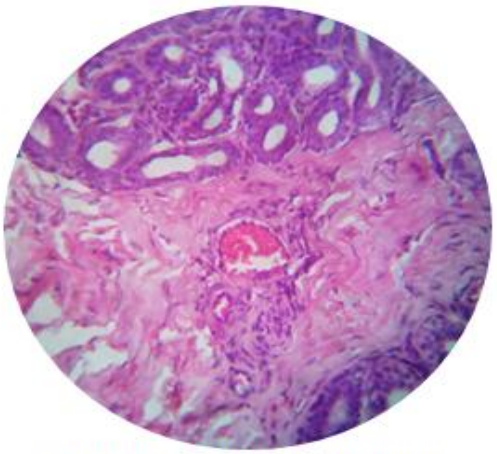

Figure 6 - Angiocentric collection of neutrophils & mononuclear cells in dermis (H&E,40x)

Hence a final diagnosis of pyodermagangrenosum was made based on fulfilling 2 major and3 minor criteria of Sue et al.

Parents were counselled, nutritional supplementation with iron, zinc and calcium. Parenteral antibiotic, saline soaks & loose dressings, topical tacrolimus and corticosteroids E/A, oral prednisolone 1mg/Kg/day was given.

Discussion

Estimated preview of PG is 3/ million population / year[3], seen more frequently in adults between 40-60 years, with female preponderance and only 4% in children. The major types of PG are classical ulcerative, pustular, bullous and vegetative.

Others are parastomal, granulomatous superficial, extracutaneous. Atypical forms are neutrophilicdermatosis of hand and pyostomatitisvegetans. Pustular form is mainly associated with inflammatory bowel disease, bullous form with hematological malignancies and vegetative form usually not associated with underlying disease.

Sue et al diagnostic criteria[2]

Major criteria:

- Rapid progression of a painful necrolytic cutaneous ulcer with an irregular; violaceous and undermined border
- Exclusion of other causes of cutaneous ulceration

Minor criteria (2/ 4)

- History suggestive of pathergy or the clinicalfinding of cribriform Scarring
- Systemic diseases known to be associated with PG
- Histopathological findings (sterile dermal neutrophilia, with or without mixed inflammation or lymphocytic vasculitis)
- Treatment response (rapid response to systemic corticosteroid)

Differences between adult and childhood form of pyodermagangrenosum (Table 1)[4]

FEATURES	ADULT	CHILDREN
Initial lesion	Maculopapular	Pustule
Common site	Legs	Face and perineum Tendency to generalize
Most frequent systemic association	Rheumatoid arthritis	Ulcerative colitis
Pathergy test	Usually positive	Frequently negative
Prognosis	Variable	Good

No laboratory test or investigation establishes diagnosis of PG with certainty. Mainstay of management is appropriate local wound care, topical and immunosuppressive therapy and treatment of underlying condition when associated with systemic diseases.

Conclusion

This case is presented for the occurrence of pyodermagangrenosum in a child which is relatively rare.

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