ATYPICAL BULLOUS PYODERMA GANGRENOSUM WITH EARLY LESIONS MIMICKING CHICKEN POX

Ramesh M[1], Kavya Raju Nayak[2], M.G. Gopal[3], Sharath Kumar B.C[4], Nandini A.S[5]

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ABSTRACT: Pyoderma Gangrenosum (PG) rare neutrophilic dermatoses (1/100,000), of which Bullous Pyoderma gangrenosum is an atypical form, which is very rare. Bullous PG is usually associated with haematological disorders like myeloproliferative disorders, haematological malignancies specially AML and several other haematological disorders. It presents as a superficial haemorrhagic bulla which ulcerates, ulcers increase in size and heal with scarring. Treatment is mainly to identify and treat the cause. Pyoderma Gangrenosum shows rapid response to oral corticosteroid therapy.[1, 3]. Clinical presentation: A 32yr old female presented with fever, multiple vesicles on face, upper limb, lower limb and trunk, and these early lesions looked like chicken pox lesions. Lesions increased in size to form haemorrhagic bullas which eroded to form ulcers, ulcers rapidly increased in size with necrotic base and erythematous to violaceous border. Investigation: Haemoglobin: 5.7gm%, Peripheral smear: normocytic and normochromic anaemia. Skin Biopsy: Sub corneal blisters with dermal and perifollicular neutrophilic infiltrate. A diagnosis of Bullous Pyoderma gangrenosum was made. Patient's anaemia was treated; oral prednisolone and topical steroids were started. Patient showed marked improvement to treatment.

INTRODUCTION: Pyoderma gangrenosum (PG) is a rare, non-infectious neutrophilic dermatoses commonly associated with underlying systemic disease. Diagnosis is based on typical clinical features and exclusion of other cutaneous ulcerating diseases. It is associated with ulcerative colitis, Crohn's disease, Rheumatoid arthritis, Connective tissue disease like SLE, certain infections and drugs. Bullous PG is a rare atypical variant, which presents with superficial haemorrhagic bulla and is usually associated with haematological disorders like haematological malignancies, myeloproliferative disorders and other haematological disorders. Skin biopsy shows intra epidermal bulla with dermal and perifollicular neutrophilic infiltrate. Treatment is to identify and treat the cause. Good clinical improvement is seen with systemic steroids and topical steroids. This article describes a rare case of Bullous PG, the early lesions of which mimicked chicken pox and patient showed marked response to systemic steroids.

CASE REPORT: A 32 year old female presented with fever, multiple vesicles and bulla on face, upper limb, lower limb and trunk, burning sensation on lesions. Many vesicles were present on an erythematous base, like a dew drop on rose petal appearance, at this point a clinical diagnosis of chicken pox was made and treated was started for the same. Vesicles increased in size to form a large haemorrhagic bulla, bulla eroded to form ulcers. The ulcers rapidly increased in size by more than 1 cm/day. These ulcers had a necrotic base and erythematous to violaceous border and central black eschar.

On examination there were multiple large ulcers present on the upper limb, lower limb and trunk, with violaceous border; floor covered with necrotic slough and central black eschar was present. The ulcers were tender, base was not indurated.

At this point differential diagnosis of Chicken pox, Bullous Pyoderma Gangrenosum, Bullous erythema Multiforme, Vasculitic Ulcer and Bullous sweets syndrome were made.

On investigation patients Haemoglobin was 5.7 gm. %, Peripheral Blood: Normocytic normochromic anaemia, Differential count and Total Leukocyte count was normal, HIV I and II: non-reactive, ANA: Negative, RA Factor: Negative. Urine routine: normal, Pathergy test was negative.

Skin Biopsy from the bulla showed subcorneal blister in epidermis, dermis showed heavy neutrophilic infiltrate along with perifollicular neutrophilic infiltrate.

With the clinical features and skin biopsy reports, final diagnosis of Atypical PG- Bullous Pyoderma Gangrenosum was made.

Treatment: Anaemia was treated with 3pint blood transfusion, patient was encouraged to take iron rich food, and Oral iron supplements were given. Wound care: Cleansing and dressing of ulcers, Escharectomy was done.

Tab. Prednisolone was given 1mg/kg body weight in divided doses; Systemic antibiotics were given after pus was sent for culture and sensitivity. Once the ulcer looked clean without necrotic tissue topical tacrolimus 0.1% and potent steroid Clobetasol propionate 0.05% was given for local application. The patient showed marked clinical improvement, ulcers size marked decreased and healed with cribriform scars.

DISCUSSION: Pyoderma gangrenosum (PG) is a rare, non-infectious neutrophilic dermatoses commonly associated with underlying systemic disease. Diagnosis is based on typical clinical features and exclusion of other cutaneous ulcerating diseases.[1]Several clinical variants has been discussed. Ahamadi and Powel classified as: Main classification: Ulcerative, Pustular, Bullous, Vegetative and Unusual presentations: Pathergic, Peristomal, Dorsal hand, Head and neck (malignant pyoderma), Multisystem, Paraneoplastic.[2]Bullous PG: This pattern has been traditionally been termed 'atypical' PG. It presents with rapidly arising, superficial, haemorrhagic bulla.[4]It shares clinical and histopathological findings with Sweet's syndrome, but typically ulcerates and heals with scarring. Bullous PG is especially associated with myeloproliferative disorder, if it occurs with IBD it is usually in patients with a significant disease.[5]

Diagnostic criteria have been proposed for the cutaneous lesions of classic ulcerative PG

Major criteria:

1. Rapid (usually >1 cm/day) progression of painful, necrolytic ulceration with an irregular, undermined, violaceous border usually with a preceding papule, pustule or bulla, and pain out of proportion to the size of the ulcerated area.
2. Exclusion of other causes of ulceration.

Minor criteria: (at least two required)

1. (a) history of pathergy, or (b) presence of cribriform scarring

2.Presence of a disease known to be associated with PG (usually listed as IBD, polyarthritis, myelodysplasia or leukaemia, monoclonal gammopathy; overall about 50% have an associated systemic disease)
3.Appropriate histopathological findings (again, specifically excluding infective causes)
4.Rapid response to oral corticosteroid therapy (usually interpreted as at least 50% reduction in size using1–2 mg/kg/day).

Histopathology: Typical findings include central necrosis and ulceration of the epidermis and dermis surrounded by an intense, acute inflammatory cell infiltrate, with a more peripheral mixed to chronic inflammatory cell infiltrate. Each clinical variant has additional, more specific, histopathological findings. In the ulcerative variant of PG, there is massive dermal–epidermal neutrophilic infiltrate with suppuration/abscess formation; in pustular PG, a perifollicular neutrophilic infiltrate with subcorneal pustule formation; the bullous variant shows a neutrophilic infiltrate with intraepidermal vesicle formation.[6] The first step in management is to search for any underlying systemic disease and treat the underlying condition. Specific treatment depends on the extent and type of disease. In limited disease, local therapy like topical or intralesional steroids or topical tacrolimus can be employed. Systemic corticosteroids are considered the drug of choice for treatment of acute, rapidly progressive form of disease. Immunosuppressant like azathioprine, cyclophosphamide, chlorambucil and colchicine has been used as adjuvants to systemic corticosteroids.

CONCLUSION: It is a very rare and interesting case, since the early lesions of bullous Pyoderma Gangrenosum mimicked that of Chicken pox. On seeing the course and with biopsy reports we came to final diagnosis of bullous pyoderma gangrenosum. Patient was put on oral prednisolone and patient's anaemia was treated. Patient showed marked clinical improvement. Hence it's important to be aware that initial lesions of bullous PG can mimic Chicken pox, and hence one should always watch for the course of disease and investigate appropriately. Once diagnosis is made, look for associated condition and treat the cause.

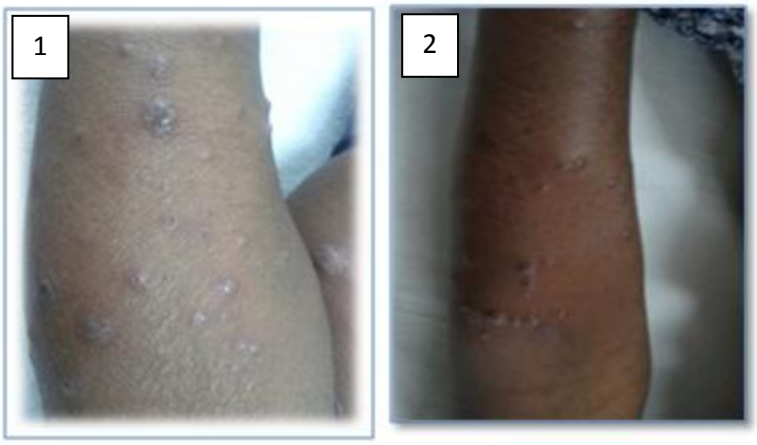

Fig. 1 and 2: Early lesions: Multiple vesicles on the upper limb mimicking chicken pox

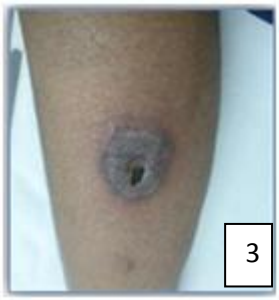

Fig. 3: Vesicles enlarged in size to form a large haemorrhagic bulla.

Fig. 4: Bulla on lower limb eroded to form large ulcers, with necrotic base, violaceous border and central black eschar.

Fig. 5: Multiple ulcers fused to form large ulcers on lower limb

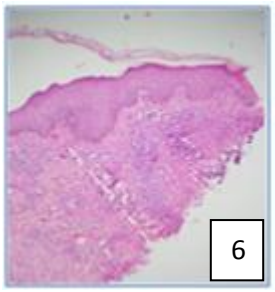

Fig. 6: Histopathology: Subcorneal blister with dense neutrophilic infiltrate in dermis

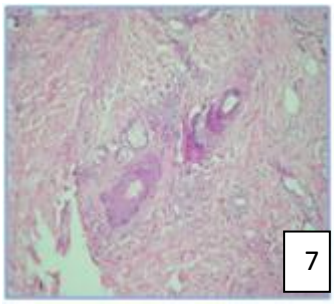

Fig. 7: Perifollicular neutrophilic infiltrate in dermis.

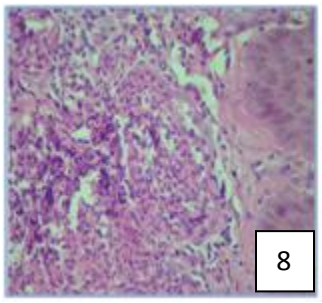

Fig. 8: Dense dermal neutrophilic infiltrate

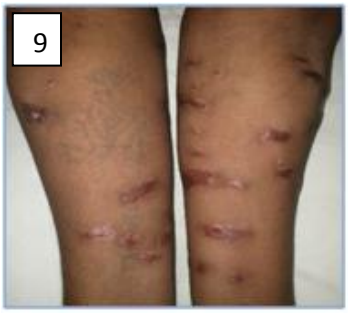

Fig. 9: After treatment ulcers on upper limb healed with cribriform scar.

Fig. 10: Ulcers on lower limb healed after treatment.

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AUTHORS:

1. Ramesh M.
2. Kavya Raju Nayak
3. M.G. Gopal
4. Sharath Kumar B.C.
5. Nandini A.S.

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Dermatology and STD, Kempegowda Institute of Medical Sciences, Bangalore.
2. Post Graduate, Department of Dermatology and STD, Kempegowda Institute of Medical Sciences, Bangalore.
3. Professor and H.O.D, Department of Dermatology and STD, Kempegowda Institute of Medical Sciences, Bangalore.
4. Professor, Department of Dermatology and STD, Kempegowda Institute of Medical Sciences, Bangalore.
5. Assistant Professor, Department of Dermatology and STD, Kempegowda Institute of Medical Sciences, Bangalore.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Kavya Raju Nayak,
71, Maruthi Layout,
Vijayanagar,
Bangalore – 560040.
Email – drkavyanayak@gmail.com

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