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Research article

A CASE REPORT ON PYODERMA GANGRENOSUM

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ABSTRACT

Pyoderma gangrenosum (PG) is a rare, destructive inflammatory skin disease which is painful and possesses rapidly progressive nature. It has life – threatening complication and devastating outcomes. The pathogenesis remains unknown and is not a bacterial infection; hence it does not possess infectious pathology but is referred to as a rare neutrophilic dermatosis with high levels of recurrent skin destruction. The disease progression can be controlled by the use of systemic corticosteroids and cytotoxic agents. Here we are reporting a case of 74 year old male patient suffering from sepsis and other comorbidities who was diagnosed with pyoderma ganrenosum by ruling out other possible causes of edematous skin lesion and purple discoloration, indicating the difficulty in the diagnosis of the condition. The patient responded to systemic corticosteroid treatment. PG causes significant morbidity to those it affects and no standardized treatment protocol exist to counteract this condition. Hence, further research is needed into the disease pathogenesis, and adequate targeted treatment.

Key Words:- Pyoderma gangrenosum, necrotizing fasciitzing, skin discoloration, neutrophilic dermatosis.

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INTRODUCTION

Pyoderma gangrenosum is a rare, destructive inflammatory skin disease in which a painful nodule or pustule breaks down to form a progressively enlarging ulcer with a raised, tender, undermined border. It was first

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described in 1916 by a French dermatologist Brocq (Brocq L, 1916). The most affected age group is from 25 to 55 years old, and this disease is more common in women. Annually, 3-10 in a million are reported as newly diagnosed cases, with 50-70% of these patients suffering from severe underlying systemic diseases; autoimmune (most frequently inflammatory bowel disease and rheumatoid arthritis), as well as hematologic disorders (leukemia and lymphoma) (Wollina U, 2007). The actiology of PG is poorly understood, although it appears to be an immune - mediated cutaneous damage. Neutrophil dysfunction, especially defects in chemotaxis, has been suggested as a possible cause of PG. It is often associated with inflammatory bowel diseases (ulcerative colitis and Crohn's disease), malignancies, arthritis and blood dyscrasias. The lesions of Pyoderma gangrenosum may begin as deep, painful nodules or as superficial, hemorrhagic pustules. Eventually these lesions develop into irregular ulcers with raised inflammatory borders and a spongy, necrotic base. It may affect any body part, although it has a predilection for the lower extremities. buttocks, abdomen, and face. The clinical presentation of PG is variable, but essentially characterized by multiple or single painful ulcerative skin lesions that progress rapidly and have a mottled, erythematous appearance (Reichrath J et al., 2005). Lesions begin as small follicular pustules or papules, which expand, becoming deep painful ulcers with well-defined, raised, over-hanging, purple bluish borders. The ulcer base is necrotic, with involvement of skin, subcutaneous tissue and muscle layers, and filled with granulation tissue, blood and purulent exudates. Reepithelialisation begins from the wound edge, healing to leave cribriform scars, which can be disfiguring, particularly if diagnosis is delayed. Patients with PG are often systemically unwell, with arthralgia, myalgia, fever and malaise. Other extracutaneous manifestations have been reported, including mucosal involvement of the oropharvnx, upper airway, eve and genitalia; sterile pulmonary, spleen, liver and bone infiltrates; and neutrophilic myositis. Atypical cases of PG occur, resembling ervthema nodosum. purpurafulminans, vasculitis. necrotising fasciitis and other neutrophilicdermatoses such Sweet's as syndrome(Alexandra T and Rachel H, 2014). Currently there exist no specific treatment for this condition, the treatment initiated depends on the case and most common prescriptions include topical as well as systemic corticosteroids, analgesics and antimicrobial agents to regulate the colonization of skin organisms in the ulcer area.

CASE REPORT

A 74 year old male patient was admitted in the surgery department with complaints of gastro fever, malaise, arthralgia, severe pain, skin discoloration and tenderness, and his frontal forearm showed edematous lesion and purple colouration. Patient is known to possess type 2 diabetes mellitus, hypertension and hyperlipoproteinemia. The medication history indicated that he was on Human actrapid, with unremarkable social history and family history. Comorbidities such as acute syndrome, sepsis, encephalopathy coronary and pancytopenia are indicated.

INVESTIGATIONS

Vital signs were normal, laboratory examination of peripheral blood smear revealed microcytic hypochromic anaemia with neutrophilic leucocytosis, haemoglobin - 8.7g/dl, total count - 28000cells/cmm, haematocrit - 19% and ESR - 46mm/hr. Provisional diagnosis was considered as necrotizing fasciitis on forearm and early septicaemia for the patient. To confirm cultures were run and came back negative for aerobic and anaerobic bacteria, hence surgical debridement was opted and biopsy was performed. Histopathologic examination showed a dense, mixed infiltrate of neutrophils,

lymphocytes and eosinophils with extensive epidermal necrosis. Based on all the presenting evidences a final diagnosis of Pyoderma gangrenosum was made. The treatment with corticosteroids: injection 500mg. methylprednisolone antimicrobial agents (combination of inj. Metronidazole 500mg, inj. Meropenem 1gm, inj. Tigecycline 100mg), analgesic; (Tramadol 1amp, paracetamol 1g) was started. The patient responded to the treatment and the stay in the hospital was uneventful.

DISCUSSION

Pyoderma gangrenosum (PG) is an inflammatory condition of the skin described in detail by Brunsting and colleagues in 1930 (Brunsting LA et al., 1930). It is characterized by the presence of deep and necrotic skin lesions with violet edges and with necrotic migratory nature, affecting any region of the body. PG has four different clinical and histopathological variants: Vegetative, bullous, ulcerative, pustular. The diagnosis of PG was based on (i) clinical findings such as painful cutaneous ulcerations with typical violaceous colour and undermined borders; (ii) histological findings consistent with PG such as dense neutrophilic or mixed infiltrate in the dermis; and (iii) exclusion of other causes of ulceration such as vasculitis, infection, vascular insufficiency or The histopathology Pyoderma panniculitis. of gangrenosum depends on the timing and site of the biopsy(Su WP et al., 1986; Clovis L et al, 2008; Mingwei J and JoshuaMingsheng Y, 2014). The clinical presentations and histopathological findings of patient were consistent with Pyoderma gangrenosum and hence the diagnosis made. The treatment with the combination of corticosteroids. analgesics and combination of antimicrobial agents is found to be effective as the lesions subsided and had completely healed over the course of the therapy. The patient also reported reduction in the pain as well as morbidity was reduced to minimum.

CONCLUSION

The condition Pyoderma gangrenosum is a painful and highly variant form of skin necrotizing disease which exposes the patient to a high degree of morbidity. The diagnosis and treatment for the same still remains empirical with no standard protocol. The aetiopathogenesis remains elusive and hence further and details investigations and research in this area is necessary to understand, diagnose as well as treat and management this condition.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors. ACKNOWLEDGEMENT Nil

CONFLICT OF INTEREST No interest

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