PYODERMA GANGRENOSUM: A CLINICAL OBSERVATION IN AN ADULT NIGERIAN

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SUMMARY

Case Definition:

Pyoderma gangrenosum (PG) is a destructive, necrotizing non–infective ulceration of the skin of unknown aetiology and rarely reported amongst Nigerians. It is believed to be a reactive inflammatory dermatosis and partly a spectrum of neutrophilic dermatoses. PG usually starts as a painful nodule or pustule that gradually becomes turgid and breaks down to form a progressively enlarging ulcer, with raised tender, undermined violeacous borders. These ulcers may occur singly or in multiples, usually with a prediliction for the lower limbs but may occur elsewhere. It is associated with several medical disorders but can also occur spontaneously in 10-30% of cases.[1] PG is a diagnosis of exclusion[2].

Objective:

The atypical presentation of Pyoderma gangrenosum in our patient from a typical hot humid tropical climate where other causes of cutaneous ulceration may simulate or mimic PG and must be excluded was the reason for revisiting this topic. A case of idiopathic pyoderma gangrenosum in a 29 years-old Nigerian male is herein discussed alongside important differentials of tropical cutaneous ulcerations that could mimic it.

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CASE REPORT

A 29-years old male student was referred to our dermatology out patient clinic with a history of a painful, advancing ulcer on the left lateral abdominal wall for three months. The lesion, which was preceded by a low-grade fever, started as a pustule that gradually broke down into the enlarging ulcer. There were no precipitating factors and no other family member had such similar complaints. He was neither a sickler, nor was he diabetic and had no history of gastro-intestinal disorders. However, he was an Igbo from an endemic zone of Buruli ulcer[3] within the region.

On clinical examination the only significant finding was a tender ulcer measuring 12cm by 7cm, on the left flank of the abdomen. The ulcer had a necrotic base with raised and undermined bluish-purple border (see fig. 1). Kobner's phenomenon was positive. He had neither oral nor genital ulcerations; neither did he have any peripheral lympadenopathy nor lymphangitis.

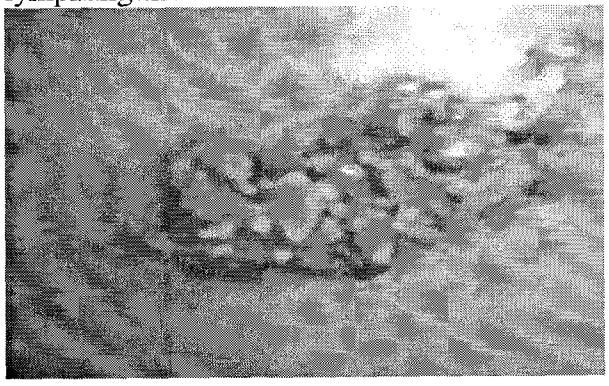

Fig 1. *Pyoderma gangrenosum exhibiting 'pathergy' on L flank of abdomen*

Routine laboratory tests revealed that the patient had slight anaemia (PCV 28%), white blood count 8,200mm^{-3}, erythrocyte sedimentation rate of 9mm/1st hr. Mantoux was negative, VDRL negative, HIV I and II were negative. Also normal were the antinuclear antibody, rheumatoid factor while the pathergy test was positive at the Mantoux site and around the lesion following minor pricks (fig.2). Liver function tests, serum urea electrolyte and creatinine, protein electrophoresis, chest X-ray, abdominal ultrasound and barium enema were all normal. Wound swab for culture was carried out 3 times and each time yielded no growth.

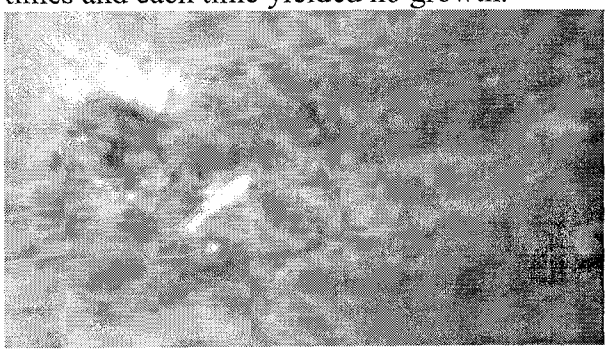

Fig. 2 *Healing Pyoderma Gangrenosum*

An elliptical incisional skin biopsy was performed and this revealed a late stage lesion of pyoderma gangrenosum with the upper dermis having a dense infiltration of lymphocytes, plasma cells and epitheloid histiocytes. Ulceration and necrosis were also noted. No atypical cells were found. Tissue cultures for bacteria, fungus and mycobacterium were negative. Pyoderma gangrenosum (idiopathic) was suspected based on the suggestive appearance of the lesions, histological features and response to therapy. These were in line with the newly proposed diagnostic criteria by Su et al from Mayo[4], used for confirming PG diagnosis.

Fig 3 *Discharging sinuses as satellite points on L abdominal flank 18 months later.*

The patient was subsequently placed on systemic antibiotics: ciprofloxacin 500mg twice daily and metronidazole 200mgs three times daily for one week, with tabs prednisolone 80mgs daily. As response to treatment was minimal he was further placed on dapsone. The ulcer was cared for daily by

cleansing with Eusol and paraffin dressing. However, he only showed up again 18 months later; after the initial four weeks of tremendous healing and improvement of his condition. This time around he had tiny sinuses oozing serosanguineous fluid around the healed areas (fig 3). Subsequently he was placed back on daily dapsone with very good response.

DISCUSSION

PG is a rare disorder and has rarely been reported amongst Nigerians. Our patient presented to the dermatology clinic as a referred case of infected ulcer of unknown origin. Being a rare disorder misdiagnosis is quite frequent as diagnosis of PG depends on recognition of the evolving clinical features. The histopathologic changes are not specific and there is no characteristic serologic or haematological marker for the disease. In addition, living in the tropics, there are several disorders/ulcers that could be confused with it. Our patient happened to come from an endemic zone of Buruli ulcer, and so it was one of our differentials. Buruli ulcers are caused by *mycobacterium ulcerans* and happen to be endemic in Africa, particularly in the Buruli county of Uganda, Liberia and Nigeria[5]. The ulcers are characteristic with undermined edges, subcutaneous necrosis and granulomas that are often, but not invariably, associated with giant cell formation. However the fact that it is painless in nature plus the clinical picture of the classical ulcerated stage gives it away[6].

Pyoderma gangrenosum is associated with systemic diseases in more than 50% of case [1, 7-9], although our patient showed no evidence of this. Some of the more common associated diseases include rheumatoid arthritis, inflammatory bowel disease, leukaemia, and acquired immunodeficiency syndrome [9-11]. Following thorough investigations our patient had none of these associations, neither did he have an abdominal stoma, but in keeping with pathergy, blunt injury[12] to the abdominal wall might have precipitated his disorder.

Ulcers of pyoderma gangrenosum may arise de novo or in areas of previous trauma, as exhibited by this patient. There are thus a couple of disorders in the tropics that could arise in a similar pattern. Although PG may occur anywhere on the body, they are unusual on the arms. Most tropical ulcers develop at a site of potential trauma, a scratch, cut, or insect bite and are therefore commonest on the legs and on the unshod foot. However, tropical ulcer is a synergistic bacterial infection that follows invasion of the skin by at least two organisms, one of which is a Fusobacterium species, usually *F. ulcerans*; the others include spirochetes or other anaerobic bacteria[13].

Cutaneous amoebiasis[14] arising from implantation/invasion of *Entamoeba histolytica* may also present like PG particularly when invasive amoebiasis escapes from the bowel of contiguous skin, usually around the anus or a colostomy, after appendicectomy, or when amoebae are implanted in another mucosa, most commonly the vagina. Amoebiasis due to *Entamoeba histolytica* is arguable the third most important human parasitic infection, after malaria and schistosomiasis in endemic area of warm humid climates.

Testing for syphilis is also important because late syphilitic ulcers may mimic PG. Moreover with the increase in prevalence of patients with HIV concurrent with syphilis in our environment[15] testing for syphilis becomes mandatory. Deep fungal infections may also mimic PG[16].

In terms of ethnicity and PG in Nigeria, Obasi[17] reported 5 cases of PG, over a 10 years period in a separate study from Northern Nigeria. All had severe and extensive PG lesions. One patient also exhibited features of malignant pyoderma gangrenosum, having had the disease for 22years with poor remission and eventually dying from the disease. Three of the patients were Hausa's, one Igbo and one, a Polynesian lady residing in Northern Nigeria. In another series from South Africa, Smith et al[18] reported two cases of PG with ulcerative colitis amongst South African blacks. The variant was the vesiculo-pustular type. All variants were similar to PG.

reported in the Western world without exhibiting any ethnic differences.

Our patient also revealed features of the vesiculo-pustular type similarly to PGs reported amongst Africans. Lesions were not confined to the lower limbs but rather to the lower left lateral flank of the abdomen, just like one of Obasi's patient. There were neither peripheral lymphadenopathy nor lymphangitis and response to treatment was one of the main pointers that helped clinch the diagnosis of PG. The addition of daily dapsone led to significant healing of this ulcer.

Indeed over the years, PG still remains a rare disorder amongst Nigerians; there is the possibility of under diagnosis in Nigeria and Africa as a whole. However, a high index of suspicion is required.

Conclusion

Appreciating features of PG observed from this idiopathic case is of utmost importance for revisiting the disorder; its features are similar to pyoderma gangrenosum described elsewhere and also the fact that ethnically there are no differences. However, misdiagnosis may be high in our environment and other tropical countries because common disorders mimicking PG are in existence. These include amoeboisis cutis, ulcernodular lesions of tertiary syphilis, cutaneous tuberculosis[19], tropical and Buruli ulcers.

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