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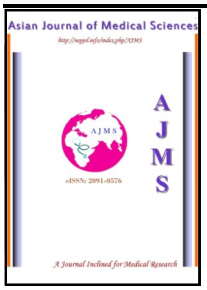


# A Rare Case of Pyoderma Gangrenosum in a HIV Positive Patient

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## Abstract

HIV infection can lead to varied spectrum of associated disease conditions. Pyoderma gangrenosum is a neutrophilic dermatosis that may be associated with myeloid malignancies. Less information is available about the association of pyoderma gangrenosum with lymphoid malignancies. We report a rare case of pyoderma gangrenosum in association with Non hodgkins lymphoma(NHL) of diffuse large B cell type. In this case the lesion which showed NHL features occurred in the perianal region, coexisting with pyoderma gangrenosum lesions in the perianal, lower limb and abdominal region. Another interesting feature is the occurrence of both these conditions in a HIV-positive patient with severe immunologic failure to first line antiretroviral therapy contributing to the refractoriness to treatment.

***Key Words:*** *HIV; pyoderma gangrenosum; Non Hodgkins Lymphoma*

## 1. Introduction

Pyoderma gangrenosum (PG) is a rare ulcerative disorder of the skin generally classified into 4 types: classic (ulcerative), bullous, pustular, and vegetative.[1] Diagnosis can be difficult, and the biopsy specimen does not provide any pathognomonic information.[2] PG is associated with many disorders including immunocompromised conditions and malignancies. Primary cutaneous B-cell lymphoma (CBCL) is a diverse group of B-cell lymphomas that are limited to the skin at time of diagnosis. The recent World Health Organization (WHO)-European Organization for Research and Treatment of Cancer (EORTC) classification of cutaneous lymphomas distinguishes 3 main types of CBCL: primary cutaneous follicle center lymphoma (FCL), primary cutaneous marginal zone lymphoma (MZL), and primary cutaneous diffuse large B-cell lymphoma.[3] Patients with Non Hodgkins Lymphoma showed cutaneous involvement in 15% to 20% of cases and in 5% to 10% of them, skin lesions are the first manifestation of the disease.[4]

Here we report a rare case of pyoderma gangrenosum (PG) with coexisting cutaneous non Hodgkins Lymphoma of diffuse large B cell lymphoma type in a HIV positive patient.

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## 2. Case Report

A 44 year old male, was found to be retropositive in 1998 and was started on antiretroviral (efavirenz, lamivudine and stavudine) therapy in 2000. Patient also had pulmonary tuberculosis 6 years back for which he was treated with WHO category 1 anti- tubercular therapy. In 2008, the patient had severe bleeding per rectum and he eventually developed a swelling over the right side of perianal region around 5 cm diameter (figure 1).

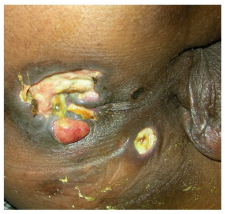

**Fig-1**

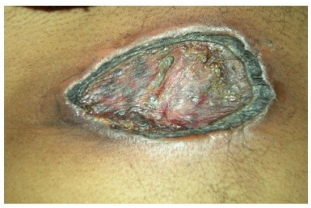

**Fig-2**

**Fig-1: A pyoderma gangrenosum ulcer and the nodule suggestive of cutaneous nonhodgkins lymphoma in the perianal region. Fig-2: ulcer with necrotic eschar and violaceous undermined margins suggestive of pyoderma gangrenous on the anterior abdominal wall below umbilicus.**

The swelling was excised and was subjected to histopathologic examination which showed diffuse large B cell lymphoma of Non hodgkins type. Following the report he was started on CHOP regimen- cyclophos-phamide, adriamycin, vincristine and prednisolone. He received 3 cycles of therapy. Since 2 months he

developed pustules and blisters followed by ulceration in the perianal region. Similar lesions occurred on right shin, right dorsum of foot and on the left side of anterior abdomen (figure 2) below the umbilicus. The ulcers rapidly increased in size with seropurulent discharge. Some ulcers had healed with necrotic centres and overhanging violaceous borders followed by peeling up of the necrotic eschars leaving raw discharging wounds again. The ulcers were painful. The discharge from ulcers was sent to us for aerobic bacterial culture and sensitivity which isolated Proteus mirabilis sensitive to cephalosporins. The biopsy of the ulcers on leg and perianal ulcer showed neutrophilic dermatosis, consisting of a diffuse inflammatory infiltrate of polymorphonuclear cells, consistent with pyoderma gangrenosum. The repeat biopsy of nodule, the remnant of the excised mass in the perianal region showed features consistent with high grade diffuse nonhodgkin's lymphoma.

Patient had severe anemia with raised ESR-130 mm/hr. FNAC of a single enlarged left side inguinal lymphnode did not show any evidence of malignancy. Ultrasound guided scan of the abdomen showed hepatomegaly, splenomegaly with multiple small ill defined hypoechoic areas with diffuse mesenteric hypertrophy probably suggestive of intra abdominal metastasis of the cutaneous lymphoma. His liver and renal function tests were normal. Lipid profile was normal except for hypertriglyceridemia 271.0 mg/dl. His CD4 count done by FACS count was 57. His viral load was 44,000 copies/ ml. So starting of second line ART regimen was planned. Regular wound management with cleaning and dressing was done.

Repeated aerobic culture and sensitivity of the seropurulent discharge was done, and, several courses of antibiotics were given as per culture and sensitivity. Cyclical CHOP regimen was with held after 4 cycles because of severe malaise, anemia and immunologic failure of the patient. The same factors were attributed to the refractoriness of the pyoderma gangrenosum ulcers to treatment. The patient was unwilling for further treatment and took to alternate system of therapy.

**3. Discussion**

The key to diagnosis of PG is excluding other causes of cutaneous ulcers through biopsy, culture, and clinical acumen. The course of PG can be acute (uniphasic), relapsing, or chronic. Relapsing or chronic courses are more likely to be associated with an underlying disease. Although the etiology of pyoderma gangrenosum is not yet clear, many reports have described associated internal disorders, including inflammatory bowel diseases (Crohn's disease, ulcerative colitis), arthritis, preleukemic states (myeloid metaplasia, refractory anemia, or polycythemia rubra vera) and myeloid leukemia.[1]

Several recent reports focus on the development of PG in immunocompromised patients.[5,6] With the increasing use of immunosuppressive therapy and chemotherapy as well as the rise of HIV infection, PG is likely to occur with increased frequency. The frequency of associated malignant disease in patients with PG is uncertain, but 7% is a reasonable estimate.[1] Leukemia, Myeloma, Waldenstrom's macroglobulinemia Polycythemia rubra vera are the various associated conditions. Myelofibrosis, lymphoma (including Hodgkin's, non-Hodgkin's[7], and cutaneous T-cell types), as well as solid tumors such as carcinoid, carcinoma of the colon, bladder, prostate, breast, bronchus, ovary, and adrenocortical carcinoma have been described in patients with PG.[1]

Pyoderma gangrenosum typically involves the lower extremities in persons without HIV infection, but the perineum is the most common site in patients with HIV infection[8] as reported in our case. Pyoderma gangrenosum lesions in HIV-infected patients are often secondarily infected with bacterial organisms.[8]

Non-Hodgkin´s lymphomas (NHL) are frequent malignancies in AIDS patients. The estimated related risk of NHL associated with HIV infection is 100 times greater than in general population, and the risk increases with the progressive immunosuppression related with retrovirus.[4] More than 90%of HIV-associated NHL is derived from B cells and the majority is high grade. Extranodal presentation is most frequent in HIV-seropositive patients than in general population. Primary cutaneous B-cell lymphomas are a heterogeneous group of B-cell extranodal lymphomas arising in the skin, without evidence of extracutaneous disease at the time of diagnosis.[9,10]

Our patient had cutaneous non hodgkins lymphoma of diffuse B cell type in the perianal region. The ulcers on all other sites were suggestive of pyoderma gangrenosum with no evidence of lymphoma. The coexistence of both the above lesions in a retropositive

case has not been reported so far to the best of our knowledge. The refractoriness of both the conditions to treatment suggests the intensity of immunologic failure in this patient.

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