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Pyoderma Gangrenosum due to Non-Hodgkin's Lymphoma: A Rare Association

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Abstract

Observation: Pyoderma gangrenosum is a necrotising ulceration of the skin with many associated diseases. Inflammatory bowel disease and myeloid leukaemias have not uncommonly been associated with this condition. Here we report an interesting but rare case of pyoderma gangrenosum due to Non-Hodgkin's lymphoma.

Introduction

'Pyoderma gangrenosum' or 'Phagedena geometrica' is an ulcerative disease of the skin whose exact cause is not known though there are many examples of depressed or abnormal immunological responses to support a belief that it is related to a defective immune system. The clinical presentation of such an ulcer is remarkable with bluish ragged margins and an unrelenting course. The etiology of pyoderma gangrenosum reveals many causes such as ulcerative colitis and also the myeloproliferative syndromes [1].

Case Report

A-46-year old male patient presented to the dermatology out patient department of our hospital with numerous subcutaneous nodules over his limbs and trunk which were present for the last 1 year, a large ulcer over his left thigh which was present for the last 2 months and intermittent fever for just over one month. The ulcer was his chief concern and it had started as small nodular lesions over his thigh, which had soon sloughed out. The patient also complained of vague joint pains for the last 6 months but did not give any history of haematemesis, melaena or hemoptysis. Cough was present for the last two weeks. There was no history of trauma at the site of ulcer nor was there any history of easy bleeding. On exami-

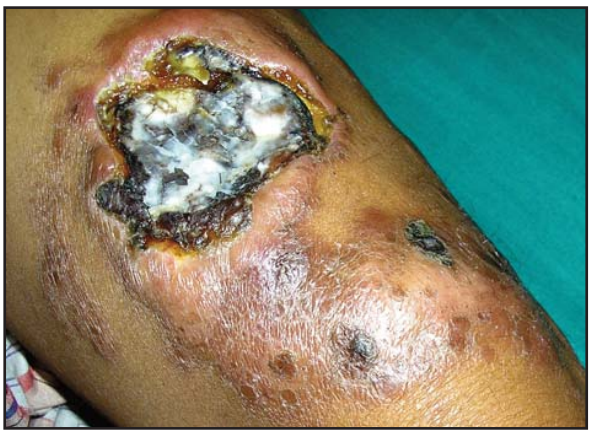

Figure 1. Large pyoderma gangrenosum ulcer on the thigh of the patient.

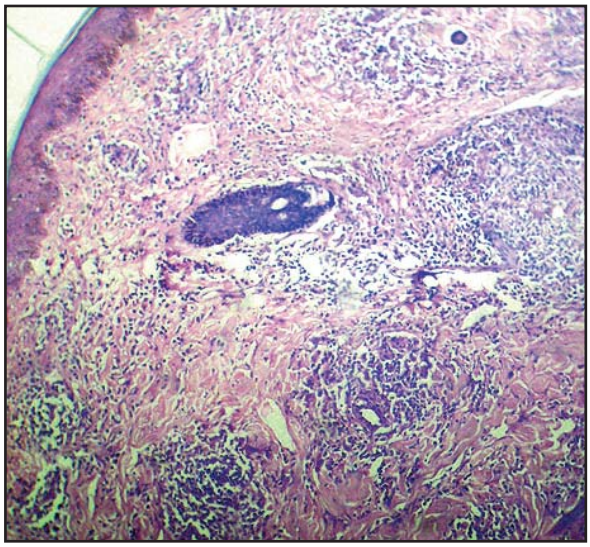

Figure 2. Histopathology showing diffuse lymphoplasmacytoid infiltrate (X10)

nation the ulcer was painless, necrotic and had a wide ragged margin (**Figure 1**). The subcutaneous nodules were nontender and there were some enlarged supraclavicular lymph nodes on both sides. He was slightly anemic, had slight jaundice but had no organomegaly. His temperature was slightly raised. Investigations revealed, WBC count: 3x109/l (N=80%, L=18%, E=1%, M=1%), E.S.R.:60 mm in first hour, Hb:7.45gm/dl, Platelets:152x109/L, Serum bilirubin:1.5mg/100ml, Serum alkaline phosphatase:576 IU/L, SGOT:132 IU/L, SGPT:60 IU/L, PPBS:160mg%, fasting sugar:121mg%. His serum urea and creatinine were within normal limits. His anti HbsAg and anti HCV antibodies were negative as were tests for HIV. His RH factor, ANF and antiphosholipid factors were negative and his chest X ray was inconclusive. Colonoscopy did not reveal any abnormality. However the FNAC (carried out from a supraclavicular lymph node of the left side) suggested a lymphoproliferative disorder with possibility of a lymphplasmacytoid lymphoma. A bone marrow examination carried out in N.R.S. Medical College with a specialized haematology unit did not reveal any lymphocytic infiltrate. A chest physician's thorough clinical examination of the Respiratory system was inconclusive and the cough was concluded to be of allergic origin. A skin biopsy was done from the margin of an ulcer and it revealed diffuse lymphplasmacytoid infiltration and epithelioid granuloma in the dermis (**Figure 2**). Finally biopsy from the subcutaneous nodules was performed and it revealed Non Hodgkin's lymphoma of high grade. Serological tests for fungal elements like sporothrix and cryptococcus were negative.

Discussion

'Pyoderma gangrenosum' manifests as a necrotic and progressive ulcer. Its exact cause has not been discovered but evidence points to a possible hypersensitivity reaction. Ample evidence of altered or depressed immunity has been found in diagnosed cases of this disease. Many conditions have been associated with pyoderma gangrenosum chief among them being ulcerative colitis and myeloproliferative disorders. However lymphoproliferative malignancies have only rarely been associated with pyoderma gangrenosum [**2**]. An enlightening case showing a $CD30^{+}$ anaplastic large cell lymphoma leading to pyoderma gangrenosum revealed increased levels of serum cytokines [**3**]. Our case presented an initial diagnostic dilemma as no cause could be found and even the bone marrow was inconclusive. Though his liver function tests revealed certain abnormalities, his antibody to HbsAg and anti HCV were within normal limits thus complicating the diagnosis. A search of the literature revealed reports of a case of 'Destructive facial T-cell lymphoma with history of nodular lesions over the abdomen and limbs [**4**]' and this set us on the right track to the association of Non-Hodgkin's lymphoma being responsible for our case. Similar to our case this case reported elevation of some liver enzyme levels but our case had no history of any intravenous drug usage as this case had. *Curtis* and *Douglas* [**5**] described a case of ulcerated pyoderma gangrenosum at the pile driver site on the arm of a person and this was found to be complicated with Non Hodgkins lymphoma. The clinical picture in many cases of pyoderma gangrenosum may confound the physician and a histopathology has been reported to give the correct diagnosis of lymphoma in a slowly growing ulcerative lesion [**2**]. Lymphomas can mimic vascular ulcers of the lower extremity and a search of the literature further revealed two cases of lower extremity pyoderma gangrenosum ulcers with T cell lymphoma[**6**].

Conclusion

We have reported our case to underline the fact that Non-Hodgkin's lymphoma can present as pyoderma gangrenosum whose thorough search of the etiology can result in an

early diagnosis, treatment and thus a favorable prognosis.

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