PYODERMA GANGRENOSUM ASSOCIATED WITH ULCERATIVE COLITIS - FIRST CASE REPORT FROM NEPAL

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A case of pyoderma gangrenosum with ulcerative colitis from Nepal is reported. The patient responded well to a combination of oral steroids, clofazimine, dapsone and salazopyrine within a period of 3 months.

Key Words : Pyoderma gangrenosum, Ulcerative Colitis

Introduction

Pyoderma gangrenosum (PG) is a rare distinctive abacterial inflammatory skin disease which was first described in 1930. Although the etiology is not known, it is associated with a systemic disease in 50% and is idiopathic in 50%.[1] It is a manifestation of altered immunity and impaired neutrophil chemotaxis. We describe a patient with PG and ulcerative colitis who responded well to multi drug therapy.

Case Report

A 20-year-old male presented with history of recurrent loose motions with blood and mucus since 5 years and multiple non healing ulcers on lower limbs since 1 year. He was investigated at Kathmandu 4 years ago where a colonoscopy and biopsy of colonic mucosa was consistent with the diagnosis of ulcerative colitis. The patient took salazopyrine for 3 years and was symptom free. However he stopped treatment and within a month

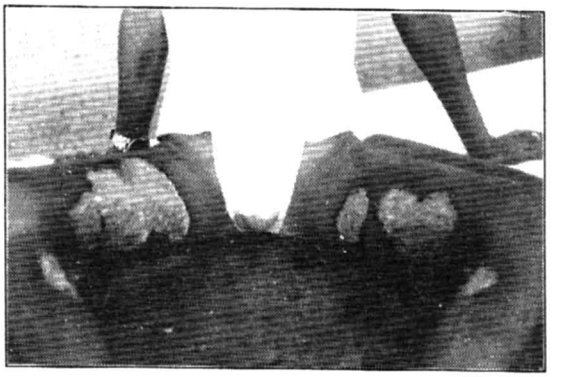

Fig.1. Multiple large ulcers before treatment

relapsed with symptoms of ulcerative colitis. He also noticed painful erythematous nodules on his right calf which ulcerated and rapidly enlarged. Few more nodules appeared on thighs which also ulcerated and remained non healing. There was no response to treatment with various antibiotics and low doses of steriods. Cutaneous examination showed multiple ulcers numbering 6, varying in size from 5-15 cm and present on medial side of thighs

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and legs. Borders were well defined, elevated, violaceous and edges were undermined. Floor was covered with

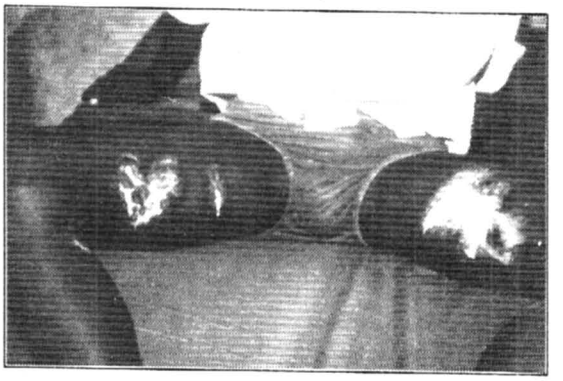

Fig.2. Complete healing after 3 months treatment

slough and bloody discharge. Few large veins were visible on the floor of the ulcers (Fig.1) Systemic examination was normal.

Investigations revealed Hb of 8.8g%, TLC-11600/cu.ml, DLC-N 69%,L 31% and ESR 51 mm in first hour. Fasting blood sugar was 76mg%. Total serum protein and serum albumin were 6.5 gm% and 2.4gm respectively. Serum Bilirubin was 0.6 mg%, SGOT - 34 IU/ml, SGPT-104 IU/ml. Mantoux and pathergy tests were negative. Rheumatoid factor was also negative. Chest X ray was normal. Pus and stool culture were sterile. Skin biopsy from the edge of the ulcer showed lymphocytic vasculitis, extravasation of RBCs and abscess formation in the dermis consistent with PG. Colonoscopy revealed red colonic mucosa with multiple diffuse ulcerations and erosions. There were multiple pseudopolyps over upper rectum and lower sigmoid colon. Mucosa was easily bleeding on touch. Colonoscope could not be inserted beyond lower sigmoid colon because of severe diffuse ulceration of mucosa. Colonic biopsy was consistent with ulcerative colitis.

Patient was admitted and started on treatment with prednisolone 20 mg.bid, ranitidine 150mg bid, salazopyrine 500 mg bid, clofazimine 100 mg bid and dapsone 100mg OD. A course of cephalexin was also given. Potassium permanganate soaks to the ulcer and dressings with soframycin were used as topical medication.

The ulcers started healing within 3 weeks of starting treatment. The dose of oral steroid was gradually tapered and all other drugs were continued in the same dosage. All the ulcers completely healed 3 months after starting the treatment (Fig.2). The patient is under regular follow-up in the OPD and is doing well on tapering dosage of all the drugs. At present he is off oral steroids and on clofazimine 200mg OD, dapsone 100 mg OD and salazopyrine 500 mg bid. He has no recurrence of either PG or ulcerative colitis after 5 months of follow-up.

Discussion

Pyoderma gangrenosum has been reported from other parts of the world and India.[2,3] Associated systemic diseases reported are ulcerative colitis, Crohn's disease, rheumatoid arthritis, seronegative arthritis, hematologic malignancies, collagen vascular diseases, monoclonal gammopathy, hepatic and pancreatic diseases, Wegener's granulomatosis and other neutrophilic dermatosis.[1] The prevalence of PG associated with ulcerative colitis has been reported to range from 30-60%.[4] Symptoms of ulcerative colitis may precede, follow or be concomitant with PG.

The treatment of PG is mostly unsatisfactory. The aim is to treat the underlying disease. Numerous modes of treatment have been tried with varying outcomes. More frequently used treatments include topical, intralesional, oral, and pulse steroid therapy, cyclophosphamide,

azathioprine, antibiotics, sulpha drugs, dapsone, clofazimine, minocycline, cyclosporine, and intensive local treatment.[5] Cyclosporine has been reported as a very effective drug for PG in the dose of 6-10 mg/kg/d with healing in 1-3 months.[1,5]

In our patient, clofazimine was given as it is known to enhance neutrophil phagocytosis and has been found to be effective in PG. Dapsone was used as a steroid sparing agent and salazopyrine was used as treatment for the underlying ulcerative colitis. Steroids were given in an adequate dosage though not as high as used in some other series.[6] Use of relatively inexpensive, safe and easily available drugs such as clofazimine and dapsone in combination in an uncommon and difficult condition such as PG which may require prolonged treatment, can be of practical importance in countries like ours.

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