PYODERMA GANGRENOSUM IN ASSOCIATION WITH ULCERATIVE COLITIS

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Summary

A case of Pyoderma Gangrenosum in a girl aged 12 years has been presented, in association with ulcerative colitis. The sites of distribution of cutaneous ulcers were typical. Literature on Pyoderma Gangrenosum has been briefly reviewed and its aetiology suggested.

The term Pyoderma Gangrenosum was originally coined by Goeckerman, Brunsting and O'Leary to describe ulcerations in association with underlying infections usually ulcerative colitis. Many years ago the French labelled these ulcers as Phagedena Geometrica. The condition is familiar to the surgeons as Meleney's undermining ulcer([1]), but the dermatologists prefer the original term Pyoderma Gangrenosum. Of late the disease has come to be regarded as a complication not only peculiar to ulcerative colitis but also other underlying wasting diseases such as empyema and rheumatoid arthritis.

The lesion may be initiated in a number of ways such as blebs, ulcers, pustules and abcesses or even a ruptured lymph node. Sometimes the patients are in robust health, but more often they have poor resistance due to serious infectious processes of long standing duration elsewhere.

Although many of the cases run a protracted course others are fulminant and end fatally. The condition is apparently more dangerous in children than in adults.

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The characteristic lesion is an ulcer, irregular in outline with a ragged, slightly raised, oedematous and boggy border usually having a distinct bluish purple colour. Beyond this border there is an erythematous areola which gradually fades into the normal skin. The central portion of the ulcer is foul and is covered with sero-purulent exudate, the removal of which reveals a clean granulating base. The margins are polycyclic or serpigenous and show the distinctive feature of being rolled under and undermined although this is not a pathognomonic sign.

The ulcers may slowly increase in size to reach a diameter of 10 cms., or more or may recede at one edge and advance at the other or may remain unchanged for a few weeks or months. There may be one or a few ulcers usually on legs, thighs, buttocks or trunk or many developing singly or in small crops at regular intervals. In Pyoderma Gangrenosum associated with ulcerative colitis the lesions tend to run a parallel course with that of the colonic changes.

Aetiology

Many different types of organisms have been isolated from lesions of Pyoderma Gangrenosum but none can

be specifically incriminated. Bacterial allergy has been postulated but not proved. Melczer proved a viral-bacterial symbiosis in which various bacteria increased the pathogenesity of the virus([2]). Rostenberg([3]) has presented the view that the disease represents a Schwartzman phenomenon. The theory that a defective immune response may be the essential factor is based on the occasional association with hypoproteinaemia or gamma .1A paraproteinaemia([4]). Since Pyoderma Gangrenosum does not respond to antibiotics and since necrotising granulomas have shown to be auto-immune reactions it may be valid to apply the concept of autoimmune reaction to this disease([2]). Malnutrition appears to be a factor sometimes. Males and females are equally affected, but majority of the cases occur in adults.

Histopathology

Histolcgical picture is nonspecific. In the region of the ulcer the epidermis is absent. The upper dermis shows necrosis with an acute inflammatory infiltrate. The mid and lower dermis shows a chronic inflammatory infiltrate. The number of blood vessels are increased with a minimal endothelial proliferation([5]).

Some authors believe that an obliterative arteritis produced by an endothelial proliferative reaction is responsible for the cutaneous lesions. Filpot et al([6]) and Mochella([7]) have described the picture of cutaneous allergic vasculitis. George Stathers et al([8]) have demonstrated the presence of giant cells around a zone of purulent inflammation but were unable to show any changes of active arteritis.

Case Report

A girl aged 12 years was admitted to the skin ward for extensive ulcerations of both the lower extremities of 20 days duration, on 28—7—72. The lesions started as boils over both the legs which in course of time bursted out resulting in small ulcers. The ulcers gradually increased in size to the extent of involving the whole of both the legs.

Patient also gave the history of having had frequent attacks of loose motions, sometimes mixed up with blood and mucous, of 5 years duration. But this was not associated with any abdominal pain.

There was a past history of similar repeated attacks of ulceration since her 6th year of age. Each attack would last for about 6–8 weeks and then heal leaving disfiguring scars. The initial lesions were noticed over the neck and upper eye-lids. The subsequent attack was at the age of 10 years when she noticed ulcerations over both the legs. After that the attacks recurred at shorter intervals.

On examination, the patient was poorly built emaciated and very anaemic. Systemic examination-NAD.

Local examination revealed extensive ulcerations over both the legs and left gluteal region.

Right Leg (Fig. 1)

Ulcer extending from the knee joint down to the ankle joint affecting the anterior surface. Size 8" x 4".

Left Leg (Fig. 1)

Two ulcers each measuring 4" x 2½" over the front of the leg.

Gluteal Region (Fig 2)

Circumscribed ulcer about 5" in diameter.

All the ulcers were covered with purulent discharge, the edges were irregular rolled and undermined and bluish

Fig. 1

Ulcers over the front of both the legs

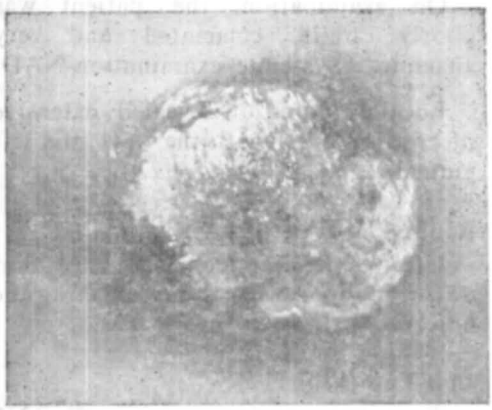

Fig. 2

Ulcer over the Left Gluteal region

in colour. Removal of the discharge from the ulcer presented granulation with a few bleeding areas here and there. The ulcers were painful and tender and the inguinal lymph nodes on both the sides were just palpable and non-tender.

Thin scars were seen over the upper eye lids, front of the neck and right knee.

Investigations

Urine Examination–NAD

(a) Macroscopic – Proglottids of T. Solium seen.

Stool Examination

(b) Microscopic–Ova of T. Solium in abundance and cyst of Giardia Intestinalis.

R.B.C. – 3.1 million/cmm.

Hb% – 8.6 gm%

Blood: W.B.C – TLC–5.150 cells/cmm.

DLC–P.64% L–32% and E–4%

E.S.R – 9 mm/1st hour.

Blood VDRL Test–Non Reactive.

Serum proteins–Total–4.4 gms%.

L.E. Cells–Negative.

Pus Culture – yielded Klebsiella aeroginosa and coagulase negative staphylococci. The organisms were sensitive to chloromyecetin, oxytetricycline, and streptomycin, but resistant to penicillin and erythromycin.

Screening of the chest – NRA.

Barium Enema (Radiologist's Report)

Barium was seen passing from the rectum upto caecum freely. No filling defect noted. Spill could be seen into the terminal ileum. The flow of Barium at the sigmoid and descending colon was rapid but the column narrowed. The normal haustrations were lost and mucosal pattern distorted (Fig. 3).

The post evacuation picture showed narrowing of the descending and sigmoid colon as well as the loss of haustrations resembling a lead pipe (Fig. 4).

Fig. 3
Barium enema picture. (before evacuation)

Fig. 4
Post evacuation picture.

These findings are consistant with ulcerative colitis.

Biopsy of the skin lesion

Showed absence of epidermis, chronic inflammatory infiltrate consisting of lymphocytes, histocytes and plasma cells in the dermis. Blood vessels were increased in number but the perivascular infiltration minimal.

Treatment given

The patient was dewormed with Mepacrine and Metronidazole. Her general condition was improved by blood transfusion followed with haematinics, proteins, anabolics and multivitamins. The treatment was with systemic corticosteroids (Prednisolone) and antibiotics (Oxytetracycline). Local therapy consisted of cleaning of the ulcers with Hydrogen peroxide and dressings with nitrofurazone.

The patient was discharged on 25–2–1973 and at the time, the ulcers were completely healed leaving depigmented scars.

Discussion

Pyoderma Gangrenosum is usually, but not always associated with an underlying bowel disturbance most often chronic ulcerative colitis. Perry and Brunsting ([9]) in their study of 19 patients of Pyoderma Gangrenosum were able to demonstrate chronic ulcerative colitis in 11 of them. Brunsting et al ([10]) have described chronic ulcerative colitis in 4 of their 5 cases of Pyoderma Gangrenosum and the fifth was associated with empyema. Samuel L. Moschella's([7]) solitary presentation was associated with an acute exacerbation of chronic ulcerative colitis. George M. Stathers et al([8]) described its association with regional enteritis. Filpot et al([6]) have attempted to relate the pathogenesis to the co-existance of diabetes mellitus and Rheumatoid arthritis in their study of three cases. The case of Pyoderma Gangrenosum reported by Nagabushanam and Patnaik([11]) was not found to be associated with any internal malady. To our knowledge this is the first case

to be reported in Indian literature with the classical picture of cutaneous ulcerations associated with ulcerative colitis.

The incidence is equal in both the sexes, majority of them being seen in adults. Our case is peculiar in that it is seen in a girl under 12 years of age.

The course of cutaneous lesions run parallel to that of the internal maladies and the general health of the patient. Our patient during her stay of 7 months in this hospital showed a remarkable improvement in her general condition, her diarrhoea subsided and finally the cutaneous ulcers healed.

The nosologic position of the disease is uncertain. Association of this condition with ulcerative colitis and the patient's response only to Corticosteroid therapy may suggest an autoimmune theory for this disease.

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