

Pyoderma gangrenosum in childhood

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Case report: Pyoderma gangrenosum is a rare painful, necrolytic skin ulcer with an irregular undermined border. It is a part of the spectrum of neutrophilic dermatoses. We report a case of pyoderma gangrenosum in a twelve year old boy. The findings reported in the literature are also discussed.

Key words: pyoderma gangrenosum, neutrophilic dermatosis, skin ulcer.

Introduction

Pyoderma gangrenosum (PG) is a painful, ulcerative cutaneous disorders of unknown etiology¹, but neutrophilic dysfunction (i.e. defect in chemotaxis or hyperactivity) has been suggested.² It begins as tender, erythematous papulosquamous or vesicle which break down rapidly to form expanding ulcers with well demarcated, undermined, violaceous borders and a surrounding areola of erythema.³ The erythema has been interpreted as an area of skin already involved but not yet showing necrosis.² Lesions arise individually or in crops on any cutaneous site, typically the lower limbs in about 75% of the cases. About 25% of the patients with pyoderma gangrenosum give history of skin trauma or pathergy in the area of lesion development, but lesion develop de novo.⁴ The ulcer can enlarge or spontaneously heal with cribriform atrophic scars. PG occurs world wide and can begin at any age, but is commonly found in the 30-50 years of age in either gender. Approximately one half of patients with PG have an associated systemic disease, most frequently inflammatory bowel disease, arthritis, hematological disease or malignancy⁴

Case report

This twelve years old boy referred to our department with history of multiple painful pustules over bilateral lower limbs, of two months duration. The lesions used to rupture to form multiple ulcers. The patient also had an associated evening rise of temperature. There was no history of similar complain in any of family members. There was no history

of significant drug intake.

On clinical examination, multiple irregular shaped crusted ulcerated lesions were seen over bilateral lower limb. The largest ulcer of 10x7 cm size was present over left shin. The margins of all the ulcers were elevated and undermined and floor was covered with unhealthy necrotic tissue. Ulcers were not fixed to underlying structures and there were no significant indurations. A few pustules were present over both legs. Physical examinations did not reveal any significant findings except for bilateral inguinal lymphadenopathy.

Laboratory investigations revealed Hemoglobin of 9.8 gm/dl, normal total white blood count and elevated ESR (46mm at end of one hour). Peripheral blood smear showed normal findings. Montoux test was negative. Syphilis and HIV serology were negative. Pus culture from the lesion was sterile. Antinuclear antibody and Rheumatoid factor were negative. Abdominal sonogram, chest X-ray and barium enema were normal. Total serum protein and albumin were within normal limits. Protein electrophoresis was not affordable to patient. Incisional biopsy from the lesion was consistent with pyoderma gangrenosum. New lesions developed in the margin of biopsy site, which was used for infiltrations of local anesthesia (pathergy test was positive). A diagnosis of pyoderma gangrenosum, idiopathic type was made based on clinical features and laboratory investigations. Patient showed recovery following treatment with prednisolone 1mg/kg body weight for 6 weeks. On follow ups no recurrence was found at 4 months.

Discussion

Pyoderma gangrenosum as such is a rare condition, which is usually seen in adults, and occurrence in children is even rarer.⁵ In a study carried out at the Mayo Clinic, in a review of 180 cases diagnosed between 1930 and 1982, it was seen that only eight cases (4%) involved children younger than 15 years of age.⁶ In a separate study of 86 patients seen at the Mayo Clinic from 1970 through 1983, four cases (4.6%) were less than 14 years of age.⁷

So far not more than 50 cases have been reported in children.⁸ Diagnosis of PG is established by a process of elimination based on suggestive appearing lesions, course, histology and response to therapy.⁵⁻⁷ PG in childhood shows certain characteristic features. As in our case, in children the lesion usually begins as pustules rather than papules or macular lesions.^{5,6}

In adults lesions are mainly seen on lower extremities⁹, whereas in children the lesions are seen on the head, neck, buttocks, perianal areas and genitals.⁵ In adults PG is most frequently associated with arthritis⁵ whereas in children it is associated with ulcerative colitis^{5,6}, followed by leukemia¹⁰ and Crohn's disease.⁶ PG has also been reported in pediatric cases with HIV infection.¹¹

However in our patient we could not detect any underlying systemic disease causing the PG.

The treatment of PG consists of mainly of use of systemic corticosteroids and sulfa drugs. Presently corticosteroids have superseded sulfa drugs as the first drug of choice. Refractory lesions have been treated with the addition of clofazamine, dapsone, rifampicine, azathioprine, cyclophosphamide and cyclosporine to steroid regimen.^{1,2} In our case patient showed recovery following prednisolone therapy for 6 weeks without any recurrence.

Patients could be rescued from aggressive debridement and even amputation by making an accurate diagnosis of PG and by providing appropriate treatment.²



Fig. 1: Characteristic pyoderma gangrenosum lesion on left lower leg



Fig. 2: Healed pyoderma gangrenosum lesion on left lower leg following 4 weeks of prednisolone therapy

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