Ind J Dermatol Venereol Leprol 1995; 61 : 96-8

PYODERMA GANGRENOSUM IN CHILDHOOD

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We have treated 4 cases of pyoderma gangrenosum in infancy and childhood in past 2 years. The ages at onset were 6 months, 8 months, 6 months and 11.5 years respectively. Initial lesions were papulopustular in 3 and nodular in 1 patients but later on all of them developed ulcerative lesions with erythematous, violaceous, infiltrated or underminded edges. The lesions were numerous in 3 patients but few in 1 patient. The first patient had very high leucocyte count. In the second patient skin lesions of PG followed measles vaccinations. We could not reveal any association in the other two patients. None of the patients responded to antibiotic therapy. Three patients were treated successfully with dapsone and one with a combination of dapsone and prednisolone. We conclude that pyoderma gangrenosum is not so rare in infancy and it is more likely to occur without associated systemic diseases.

Key Word : Pyoderma gangrenosum

Introduction

Pyoderma gangrenosum (PG) is a skin disorder of uncertain origin, characterized clinically by the presence of one or more chronic cutaneous ulcers with characteristic undermined purple or red margins and halo of surrounding erythema. The primary lesions is a pustule, papulovesicle, nodule or blister . The necrotising inflammatory process extends peripherally form the primary lesion, resulting in ulcer formation. Lesions are often initiated by trauma. It commonly occurs in middle aged patients but is rarely seen in childhood. We have treated 4 cases of PG in infancy and childhood in last 2 years. The data of these patients are reported.

Case Reports

Case I : A boy aged 8 months was having multiple ulcerative lesions of different sizes ranging from 1cm to 10cm on various sites of the body with erythematous violaceous infiltrated margins and necrotic slough in the floor for 2 months. Initially the lesions were small papulopustules, which ulcerated within 2-3 days and gradually enlarged in size. During hospitalization he developed a few new lesions at injection sites. The patients was febrile.

Repeated pus culture and blood culture did not reveal growth of any pathogenic organism, Complete blood count showed a leucocytosis (total count 39,000/cu mm with segmented neutrophils 25%, band neutrophils 43%, lymphocytes 24%, monocytes 5%, eosinophils 1%, metamyelocyte 1%) with thrombocytosis (822,000/cu mm). Blood tests for urea, creatinine, electrolytes, liver function tests, sugar, VDRL, serum electrophoresis for proteins and immunoelectrophoresis did not reveal any abnormality. Routine examination of urine and stools also did not show any abnormality. Slit smear from the infiltrated margin of an ulcer stained with Giemsa stain did not show any L.D. bodies. X-ray chest and ultrasound of abdomen also did not reveal any pathology.

Biopsy showed absence of epidermis and infiltration of upper dermis with large number of degenerated neutrophils. There were large numbers of newly formed thin-walled blood vessels. The infiltrate had extended through the dermis to the subcutaneous tissue in a diffuse manner.

Initially the patients was treated with high

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doses of antibiotics - ampiclox, gentamicin and cefotaxime without any significant improvement. Then he was given oral prednisolone 15-25 mg/day and showed good response within one week. Later he started developing new lesions so the treatment was supplemented with dapsone 12.5 mg/day. The lesions healed almost completely within 6 weeks. The dose of prednisolone was gradually tapered to 5 mg/day but dapsone was continued in the same dose for further 6 weeks. The lesions healed completely with atrophic scarring. The patients returned with a recurrence of a few lesions after 3 months and was treated successfully with prednisolone and dapsone.

Case 2 : A 9-month-old baby girl presented with multiple papulo-pustular lesions of 3 weeks duration, which soon ulcerated and increased in size. The patients had multiple deep ulcers with undermined edges on her back, chest and scalp. Pus culture revealed growth of Escherichia coli and Pseudomonas aeruginosa. Other investigations did not reveal any abnormality. The patients was vaccinated against measles one week before the onset of skin lesions.

The patient was initially treated with broad spectrum antibiotics for 3 weeks with partial improvement and was discharged. After one month the patient returned with a recurrence of lesions. This time pus culture did not show any growth of micro-organism. Biopsy taken from the margin of the lesion showed necrosis of the epidermis with infiltration of neutrophils in the dermis and subcutaneous tissue. This time the patients did not show any improvement with systemic antibiotics, so she was given dapsone 12.5mg/day which showed dramatic response within one week and the lesions healed completely within 4 weeks.

Case 3 : This girl aged 2 years came with history of developing nodulo-ulcerative lesions, one after the other on various sites of the body since the age of 6 months. At the time of admission the patient had an ulcerated lesions 3cmx3cm, with undermined violaceous margins on the chest. There were atrophic scars of old lesions on the left leg, right upper arm and neck. Histopathology from the margin of the active lesions showed similar changes like patient number 2. Other investigations did not show any appreciable abnormality.

The patient was initially treated with local and systemic antibiotics for 3 weeks without any significant improvement. Then the patient was given dapsone 25 mg/day and the lesions healed completely within 3 weeks. Dapsone was discontinued after 4 weeks. She came with recurrence after 4 months with a similar lesion on the right thigh. Dapsone was again started and lesion soon healed but dapsone was continued for further 2 months with tapering dose. The patient did not have any recurrence after that.

Case 4 : A girl aged 12 years presented with multiple well-defined necrotic ulcerated lesion with infiltrated margins on the knees, elbows and buttocks for 6 months. Lesions initially appeared in the form of papulopustules which soon ruptured to develop ulcerative lesions with gradual extension. Pus culture showed growth of Staphylococcus aureus. Other investigations on blood, urine and stools, X-ray chest and ultrasound abdomen did not reveal any significant abnormality. A Mantoux test was also within normal range.

Biopsy revealed neutrophilic infiltration of the subepidermal region with a heavy chronic inflammatory infiltrate of the lower dermis extending deeply into subcutis. There

was evidence of hyperplasia of the epidermis at the edges of the ulcer. The patient was first treated with local and systemic antibiotics with control of secondary bacterial infection, but the size of the ulcers was increasing and a few new lesions developed. She was given dapsone 100 mg/day, and showed a dramatic response within one week and her lesions healed completely in one month. Dapsone was continued in a dose of 50 mg/day for one month. This patient came with recurrence of few lesions after 3 months. This time also she was successfully treated with dapsone.

Discussion

The age at onset in patient number 1 and 3 was 6 months. To our knowledge, PG in this age has not been reported earlier, although Glass et[1] have reported a case of PG in a 7 months infant with clinical features similar to our case number 1.

Except patient number 3 all of our patients had numerous lesions while in previously reported cases, lesions were few.[2]

Development of new lesions at the injection sites in case number 1 and presence of lesions mainly on the pressure sites in patient number 4 suggested the phenomenon of pathergy as also recorded in previous reports.[2]

Patient number 2 was vaccinated against measles, one week before onset of the lesions. Similar causal relationship of different vaccines to PG has been reported before.[1] We could not reveal any associated disease in the rest of the patients, however, further follow-up is needed. In general PG is mainly associated with ulcerative colitis, Crohn's disease, rheumatoid arthritis, monoclonal gammopathy, varied myeloproliferative disorders, malignancies and various immnuological disorders. In 50% of the cases, PG occurs in absence of any associated diseases.[3]

All of our patients had a chronic course of the disease and had recurrences. None of them responded satisfactorily to antibiotic therapy. The first patient responded well to a combination of prednisolone and dapsone. Glass et al[1] have also observed good results with similar combination therapy. The rest of the patients responded well to dapsone alone.

We conclude that PG in infancy is not rare and it is more likely to occur without associated systemic disease.

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