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Pyoderma gangrenosum in Infancy

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Abstract. A case of pyoderma gangrenosum is described in a patient aged 9 months. The histopathological features were those of acute panniculitis and treatment with oral prednisolone produced an excellent clinical response. No underlying cause of the skin lesions has been found. The differential diagnosis of the clinical and histological appearances seen in our patient is discussed.

Key words: Pyoderma gangrenosum; Panniculitis

Pyoderma gangrenosum is an uncommon ulcerative skin disorder that has not, as far as we are aware, been reported previously in early infancy. We wish to report a case with somewhat unusual histological features in a 9-month-old infant.

CASE REPORT

The patient, a previously healthy female infant, developed an upper respiratory tract infection accompanied by cervical lymphadenopathy at the age of 9 months. She was treated with ampicillin, with initial improvement, but a week after the onset of her illness she began to

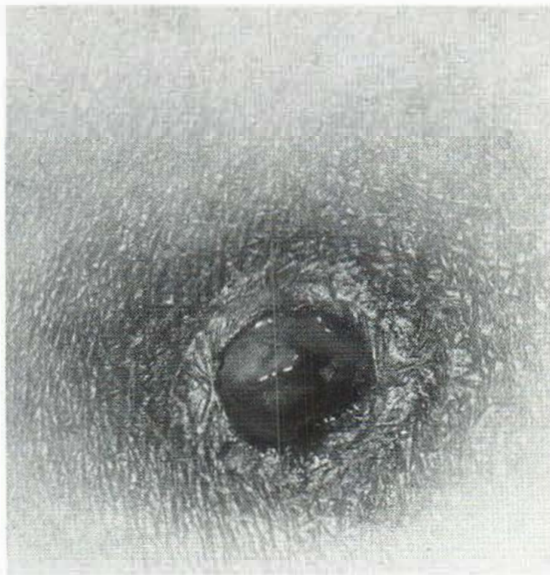


Fig. 1. Fully developed lesion on the leg, showing pustulation and ulceration.

refuse feeds and an erythematous lesion was noticed on her left cheek. Treatment was changed to topical fusidic acid ointment and oral amoxycillin, but more lesions appeared followed by swelling of her right foot and she was admitted to hospital.

On admission she appeared ill, with a temperature of 38.3°C, pustular lesions on her face, and a swollen right foot. She was otherwise a well-nourished child whose length and weight were both around the 50th centile. She had no lymphadenopathy, her throat and ears appeared normal, and examination of other systems proved essentially negative. She was found to be anemic, with a haemoglobin of 9.0 g/dl (normochromic, normocytic) and had a pronounced neutrophil leukocytosis (total white cell count 27.8×10^9 /dl, 80% polymorphs). Her ESR (Westergren method) was 110 mm in the first hour. An X-ray examination of her right foot and ankle revealed no abnormality and a chest X-ray was normal.

She was initially thought to be suffering from a bacterial infection and was treated with benzyl penicillin and flucloxacillin. Bacterial cultures from the pustules and blood were repeatedly negative. After 5 days in hospital she still had an elevated temperature and was continuing to develop fresh skin lesions on her legs, on her left arm at the site of a recent Mantoux test and at the sites of injections on her thighs. Her treatment was changed to intravenous gentamicin and flucloxacillin. The skin lesions began as small erythematous papules that developed into large pustular ulcers over a period of 36 hours and she had, by this time, ulcers on her left thigh, calf and arm, as well as her right arm, and right leg (Fig. 1). The haemoglobin concentration fell further to 7.7 g/dl and she was transfused with whole blood. The total white cell count rose to a maximum value of 41.9×10^9 /dl.

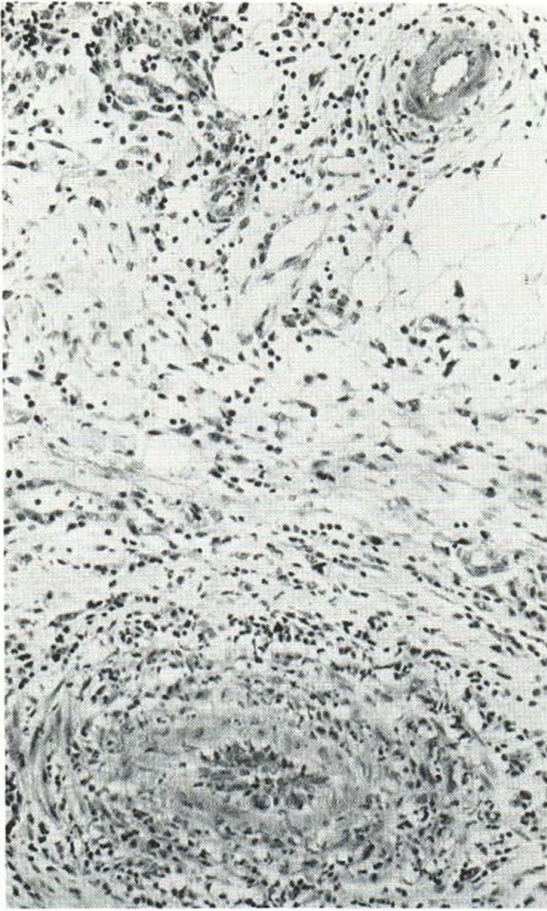


Fig. 2. The histological features of an early lesion, with acute panniculitis and endothelial proliferation in larger blood vessels. Original magnification, $\times 30$.

The histological features of an early, non-ulcerated lesion of the left thigh were those of acute panniculitis with inflammatory exudate and obliterative endothelial proliferation of larger blood vessels (Fig. 2). There was no histological evidence of a vasculitis as such and the overlying dermis and epidermis were normal.

The following investigations proved abnormal. Serum IgM was elevated at 170 mg% (normal 31–149 mg%). The rheumatoid factor was weakly positive. A defect of polymorph chemotaxis towards casein and activated plasma was detected. The electrocardiograph showed sinus rhythm with a minor degree of right bundle branch block on one occasion but normal conduction on subsequent occasions, and there was no evidence of ventricular hypertrophy.

The remaining investigations gave normal or negative results. These included repeated bacteriological cultures of skin, blood, urine and faeces, as well as viral culture and serology.

In view of the negative bacteriological findings, the

poor response to antibiotics, and the demonstration of an inflammatory process involving subcutaneous blood vessels on skin histology, the patient was commenced on treatment with prednisolone in an initial dose of 20 mg daily. The response to prednisolone was dramatic. The temperature never exceeded 37.4° after the first dose of prednisolone, whereas temperatures of over 38° were recorded on most days before steroid treatment was begun (Fig. 3). The ulcerated lesions began to heal and no fresh skin lesions developed. After 2 weeks the dose of prednisolone was reduced and after 3 weeks the patient could be discharged from hospital with her skin lesions completely healed. Nine months from the onset of her symptoms she remains well and the prednisolone has been discontinued.

DISCUSSION

The unusual features of this case of pyoderma gangrenosum are the patient's very young age of 9 months and the histological changes in the skin. The absence of an associated disease is not particularly unusual—8 patients in the series of 15 cases of pyoderma gangrenosum reported by Hickman & Lazarus (6) had no associated systemic disease. It seems likely that the condition is very rare in infancy—at least in the acute, progressive form. The clinical features are distinctive once the possibility of an infective pyoderma has been excluded. In adults, the disease is said to pursue either an acute progressive course with rapid evolution of new lesions until the disease is halted by treatment, or a more chronic indolent course (7). In our patient there was no tendency to improve until corticosteroids were administered, whereupon there was a very prompt and unmistakable clinical improvement that has subsequently been maintained.

The commonest histological finding in pyoderma gangrenosum is an area of ulceration with a dense inflammatory infiltrate of predominantly neutrophil polymorphs throughout the dermis (1). There have been reports of vasculitis (4) and the role of vascular changes in the early development of the lesions has not been resolved. Immunofluorescence studies to date have been negative (8). The presence in our patient of an inflammatory infiltrate in the wall of a medium-sized subcutaneous vessel raised the possibility of polyarteritis nodosa. Infantile polyarteritis nodosa usually has an extremely grave prognosis (10) although it has been suggested recently that there is a more benign form of the disease represented by the mucocutaneous lymph node syndrome (2). Our case bore no clinical resemblance to either of these disorders. A purely cutaneous form

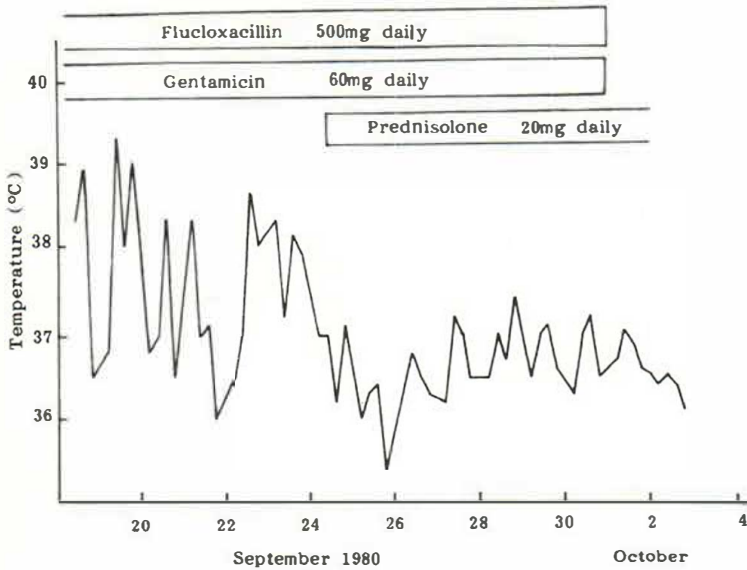


Fig. 3. Temperature chart showing an immediate response to the administration of prednisolone and the lack of effect of antibiotics.

of polyarteritis nodosa has been described in adults (3, 5) and in a child (11). This form of the disease has a much better prognosis than the systemic variety and is characterized by cutaneous nodules, ulceration and livedo reticularis. Our patient's lesions showed little sign of a nodular phase and there was no livedo. The presence, also, of lesions at the site of injections or skin trauma, as in this case, has been commented upon in previous reports of pyoderma gangrenosum (9) but not in polyarteritis nodosa.

The discovery of a defect in polymorph function is interesting, as a similar finding has been reported before in pyoderma gangrenosum (8) but the interpretation is fraught with difficulty as polymorph function can be affected by a variety of factors including drug treatment, and methodological variables (12). While there is some evidence to suggest that pyoderma gangrenosum is an immunologically mediated disease, the precise mechanism of pathogenesis remains unknown.

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