Pyoderma gangrenosum in a 2 ½ year old boy

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Abstract

A 2 ½ year old boy from Yatiyanthota presented with rapidly spreading leg ulceration of one month's duration. The large ulcer involving most part of the right calf had a characteristic violaceous undermined edge surrounded by a peripheral zone of erythema. A diagnosis of Pyoderma gangrenosum was supported by a dense dermal neutrophilic infiltrate with few giant cells on histology. The condition improved dramatically to systemic steroids. Extensive investigations did not reveal any known associated disease.

Introduction

Pyoderma gangrenosum is an uncommon chronic painful ulcerating skin disease occurring mainly in the lower limbs of adults. The lesions have a distinct clinical appearance of necrotic ulceration, mucopurulent base and a violaceous undermined border surrounded by a peripheral zone of erythema. Pyoderma gangrenosum does not respond to antibacterial agents even when parenterally administrated. Reports of this disease in children are rare.

Case report

A 2 ½ year old boy was referred from a paediatric surgical unit to the dermatology unit at LRH with a rapidly spreading non-healing painful ulcer affecting the right leg. The ulcer followed a leech bite, but has progressed rapidly both in extent and depth since then.

Figure 1.

This is despite many courses of broad-spectrum intravenous antibiotics based on culture reports and several wound toilets carried out under general anesthesia. At the time of the referral ulceration involved most part of the right calf and adjacent lower thigh. Prior to this child has not suffered from any major illnesses.

On examination the child was febrile and was in severe pain. A large oval shaped ulcer was found or the posterior medial aspect of right leg and adjacent lower thigh. The edge of the ulcer was violaceaous, undermined and surrounded by a peripheral zone of erythema. The base was relatively fresh with new granulation tissue. System examination was normal. In particular there was no adenopathy or hepatosplenomegaly.

An initial investigations showed ESR - 91 mm, a white cell count of 24,000/mm with a normal differential and a left shift. Liver/Renal profiles, x-ray chest, abdominal ultrasonography were all normal. An immunological profile made up of immunoglobulin levels, lymphocyte subset, neutrophil fraction tests ANA, and Rheumatoid factor were all normal.

Histology from the wound edge showed a mixed inflammatory infiltrate with dermal neutrophilic abscess formation, a lymphohistiocytic infiltrate with giant cells. Blood vessels showed a mild lymphocytic vasculitis with capillary and venous thrombosis and red blood cell extravasation.

A sigmoidoscopy was performed to exclude associated inflammatory bowel disease. Macroscopically colonic mucosa was normal as was the histology of random biopsies of the colonic mucosa.

Management

Patient was started on hydrocortisone 50 mg 6 hourly intravenously. Within 24 hours of initiation of treatment marked improvement of the patient was noticed. After 48 hours IV Hydrocortisone was stopped and 10mg of oral prednisolone was given daily. On this the ulcer continued to heal rapidly and was complete by one week with a typical cribriform

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scar. (See Figure 2) Presently the child is on prednisolone 5 mg every other day.

Figure 2.

Discussion

Pyoderma gangrenosum is an uncommon cause of cutaneous ulceration in children. As such it is often not recognized, at least initially, in clinical practice. The condition is a cause for severe morbidity and does not respond to antibacterials even when parentrally administrated. The later merely adding to the cost. The lesions have distinct clinical appearance of necrotic ulceration, a muco-purulent base and a very characteristic violaceous undermine border surrounded by a peropheral zone of erythema. In our patient ulcer base was relatively clean possibly due to the repeated use of broad spectrum IV antibiotics. Rarely Pyoderma gangrenosum can present as bullous, pustular, vegetative lesions. The lesions are often found on the lower limbs and often proceeded by minor trauma, a phenomenon termed pathergy. The leech bite in our patient explains this well. Histopathology of this condition is non specific and vary depending on the age of the lesion and the site of the biopsy. Therefore the diagnosis remains clinical.

In the management of this condition in children it is important to evaluate the child for possible associated systemic illnesses. This include inflammatory bowel disease, arthritis, haematological malignancies and immunodeficiency. All these were reasonably excluded in our patient. Exclusion of immunodeficiency is very important as the main stay of treatment is corticosteroids. Other treatment options include Dapsone, sulphasalazine and cytotoxic agents, all with potentially severe side effects in children. Topical Tacrolimus is an affective agent for this condition. However this is expensive and is not freely available. It is important to follow these patients as Pyoderma gangrenosum can predate the underlying systemic disorder.

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