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Case Report

Childhood pyoderma gangrenosum: diagnosis often missed

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ABSTRACT

We present a 10-year-old male child who presented with 2 months history of fever and extensive ulceration over both lower limbs and buttocks. The new lesions were bullous and pustular, so working diagnosis of bullous impetigo was considered. WBC count showed mild anemia and neutrophilic leucocytosis. Peripheral smear showed neutrophilia, adequate platelets and normal lymphocytes. Blood culture and pus culture were sterile. Stool for occult blood was negative, liver function tests were normal. Mantoux test was negative. Serum immunoglobulins were normal and HIV, HBsAg tested negative. Skin biopsy showed foci of granulomatous inflammation with neutrophilic infiltration. With persistent neutrophilia and neutrophilic infiltration of ulcers, pyoderma gangrenosum was kept as diagnosis. The child was started on local steroid application and oral prednisolone 1 mg/kg/day. Wthin 2 days of oral steroids child became afebrile. Ulcers started healing and neutrophila also normalized. In view of reappearance of fever, immunosuppressive, closporin was started, after which child responded well and ulcers healed completely. So, in conclusion, we report a paediatric case of pyoderma gangrenosum which poses diagnostic difficulty, particularly to non-dermatologists. The dramatic response to steroid and cyclosporine helped cure the ulcers and remit the disease and sufferings.

Keywords: Chronic skin ulceration, Pyoderma gangrenosum

INTRODUCTION

Childhood pyoderma gangrenosum is a rare cause of cutaneous ulceration in Pediatric age group, accounting for 4% cases of pyoderma worldwide.[1] Distinctive cutaneous ulceration which is usually idiopathic but may be associated with many systemic disorders. The etiopathogenetic of PG (pyoderma gangrenosum) is still not well understood.

Clinically it is classified into ulcerative, pustular, bullous and vegetative types. Characteristic phenomenon of 'pathergy' is seen in pyoderma gangrenosum in which superficial trauma to the skin can cause ulceration. Dramatic response to steroids and other immune-suppressants is seen.

CASE REPORT

We present a 10-year-old male child who presented with 2 months history of fever and extensive ulceration over both lower limbs and buttocks. Fever was high grade and ulceration was deep and showed pus (Figure 1, 2 and 3).

There was no organomegaly, lymphadenopathy. Respiratory, cardiovascular and central nervous system examination was unremarkable. The new lesions were bullous and pustular, so working diagnosis of bullous impetigo was considered. WBC count showed mild anemia and neutrophilic leucocytosis (Hb-11 gm% and WBC count: 26000/cmm with 80% neutrophils and normal platelet count). Skin biopsy showed foci of granulomatous inflammation with neutrophilic

infiltration. Granuloma consists of lymphocytes, plasma cells, histiocytes occasional langhan cells and foreign body giant cells. Overlying epidermis showed psoriasiform changes and neutrophilic infiltration. Initially treated with IV amoxycillin. Blood culture and pus culture were sterile.

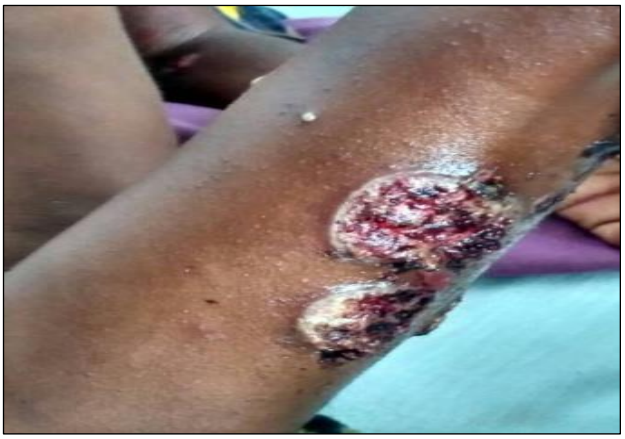

Figure 1: Large ulcer with undermined edges.

Figure 2: Large ulcer on anterior aspect of right tibia.

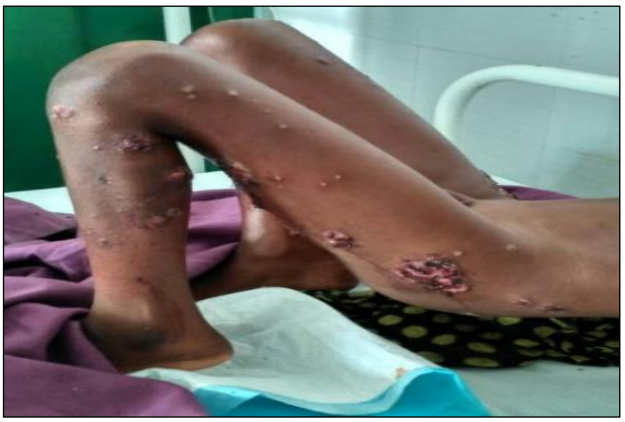

Figure 3: Pustules formation on superficial trauma-phenomenon of pathergy.

Child continued to have fever. Though old ulcers started healing, new pustules kept on emerging on site of trauma and kept ulcerating. CBC showed persistent neutrophilia. Hematologist opinion taken and bone marrow biopsy was planned but patient did not give consent for the same. With persistent neutrophilia and neutrophilic infiltration of ulcers, pyoderma gangrenosum was kept as diagnosis. Peripheral smear showed neutrophilia, adequate platelets and normal lymphocytes. Stool for occult blood was negative, liver function tests were normal. Mantoux test was negative. Serum immunoglobulins were normal and HIV, HBsAg tested negative. The child was started on local steroid application and oral prednisolone 1 mg/kg/day. Within 2 days of oral steroids child became afebrile. Ulcers started healing and neutrophilia also normalized. After 7 days of steroid, there was much improvement in ulcer healing, but fever and neutrophilia started again. So steroid dose was increased to 2 mg/kg/day. In spite of increasing the steroid dose patient continued to have fever for 5-7 days after which oral cyclosporine A was started at the dose of 5 mg/kg/day. Patient became afebrile in next two days and was discharged home on cyclosporine A after 5 days. Patient regularly followed up in paediatric OPD. Steroid was tapered and stopped over next 2 weeks. Cyclosporine was continued for 2 months and then gradually tapered over 1 month and stopped. CBC and Liver function tests were monitored every month and were normal. Patient's ulcers healed completely So we labelled this patient as idiopathic pyoderma gangrenosum and child was ambulatory, with superficial scars on legs (Figure 4). Lesions did not appear again.

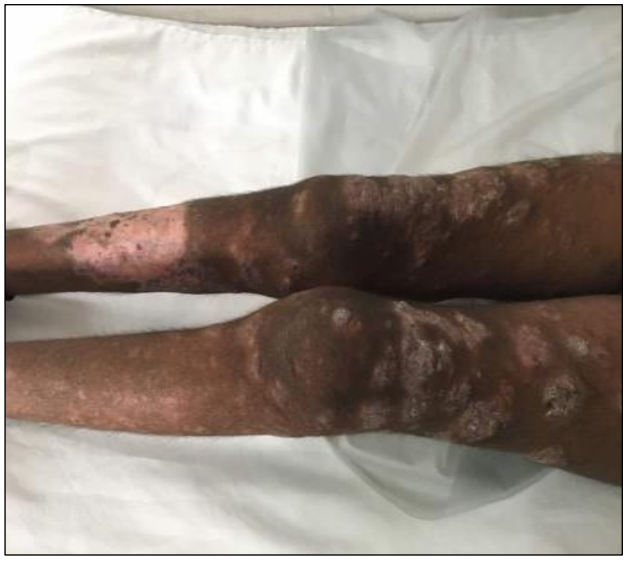

Figure 4: Healed ulcers after chemotherapy.

DISCUSSION

Pyoderma gangrenosum (PG) was first described in 1930 by Brusting and colleagues.[2] PG is rare in infants and children with an estimated incidence of about 4% pyoderma gangrenosum lesions start as pustule and rupture to form ulcers.[1] These ulcers would enlarge, have undermined margins with superficial gangrenous

changes. There is pathergy phenomenon in which even a superficial trauma to the skin will cause ulceration. So surgical treatment would harm in such a scenario. Diagnosis is clinical and depends on the exclusion of other causes of cutaneous ulceration. No specific pathologic or laboratory findings exist. Concurrent systemic disease occurs in 50% of affected patients, remaining cases are thought to be idiopathic or autoimmune.[3,4] Associated systemic diseases include inflammatory bowel diseases, rheumatoid arthritis, SLE, hematological malignancies, chronic active hepatitis, congenital or acquired hypogammaglobulinemia.[5]

The lesions of PG have been classified into 4 types depending on clinical and histological presentation.[4] 'ulcerative' is the commonest type in which lesions are most commonly located on lower limbs but may occur anywhere. Lesions often begin as pustules but can also be pathergic in etiology. A large ulcer with a well-defined, undermined border surrounded by a halo of erythema is usually formed within a few days at the site of minor trauma. Pustular PG, although all PG lesions start as pustules, sometimes they do not progress to ulceration. Third type is bullous in which they begin as bullae and are superficial. Patients with this type of PG develop painful rapidly enlarging bullous lesion which becomes superficially erosive and then ulcerative, especially on the dorsum of the hands.

It has been proposed that atypical PG and sweet's disease are points on a continuum.[6] Forth type is vegetative PG, also known as superficial granulomatous pyoderma, presents as chronic, non-progressive, superficial ulcer that is not painful and often lacks a violaceous undermined border.[4] The diagnosis of PG is established by consideration of clinical features, review of histopathologic findings and with the benefit of negative culture results and other investigations. Histopathological findings in PG are not specific. However, a biopsy is suggested in all instances to exclude other diseases. Biopsy should not be deferred because of concerns of pathergy. Microscopic features include extensive sterile dermal neutrophilia.

Management of PG is divided into local wound care, topical therapy and systemic therapy.[7] The treatment of underlying disease may aid in healing. In patients without an identifiable associated disease, it is still possible for it to appear later; hence follow-up and evaluation are required even after the skin lesions have healed.[8]

In the case of sloughy or purulent ulcers wet compresses with saline and alginate dressings are useful. Although some topical agents such as tacrolimus, potent corticosteroids, and cyclosporine have reported efficacy, evidence from large clinical trials is lacking.[9] Systemic corticosteroids have been the most predictable and effective treatment of acute, rapidly progressive form of the disease. High doses of prednisolone or pulse therapy with methylprednisolone/dexamethasone may have to be used in resistant disease.[10] Among the immunosuppressive agents, cyclosporine, which does not cause significant myelosuppression has proved to be a useful substitute therapy for PG resistant to steroid treatment.[8]

Cyclosporine is considered as most effective steroid sparing agent. In steroid resistant cases of PG, combination of corticosteroids with cyclosporine or azathioprine are considered as first choice therapy on the basis of clinical experiences though there are currently no general recommendations for the management of PG. Other alternative options are mycophenolate mofetil, dapsone, sulfasalazine, cyclophosphamide and biologics. Tumor necrosis factor alpha (TNF-alpha) blockers and other injectable biologics have been demonstrated to be successful.[11] Infliximab (5 mg/kg/week intravenously at weeks 0, 2, 6 and at every 6-8 weeks), Adalimumab (40 mg subcutaneously weekly), all seem to be effective in PG especially in association with IBD.[12]

CONCLUSION

So, in conclusion, we report a paediatric case of pyoderma gangrenosum which poses diagnostic difficulty, particularly to non-dermatologists. The dramatic response to steroid and cyclosporine helped cure the ulcers and remit the disease and sufferings.

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