CASE REPORT

Pyoderma Gangrenosum with Pure Red Cell Aplasia

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Pyoderma gangrenosum is an inflammatory condition of the skin commonly associated with inflammatory bowel disease and rheumatoid arthritis, but also associated with various hematological malignancies. We describe its association with pure red cell aplasia in a four year old boy who presented with progressive skin lesions, fever and pallor, and improved with corticosteroid therapy.

Key words: *Pyoderma gangrenosum, Pure red cell aplasia, Superficial granulomatous pyoderma.*

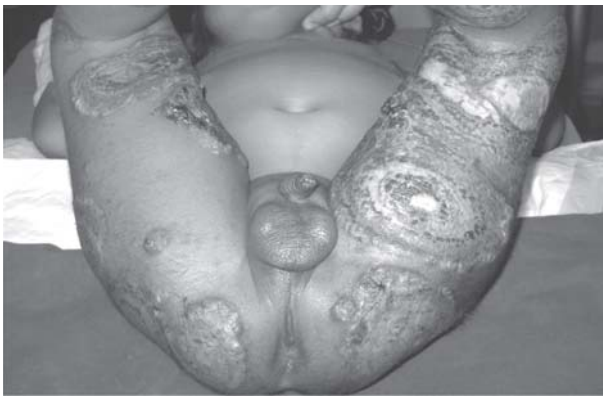
Pyoderma gangrenosum (PG) is a rare, noninfectious neutrophilic dermatosis, commonly associated with inflammatory bowel disease and rheumatoid arthritis and also rarely with myelo-proliferative disorders and hematological malignancies. The diagnosis of PG is clinical, along with compatible histological findings. To our knowledge there is no report of association of pyoderma gangrenosum with pure red cell aplasia in children.

CASE REPORT

A 4-year old boy, 1st born of non-consanguineous parentage, who had normal developmental milestones, presented with complaints of painful progressive skin lesions involving the buttocks and both thighs since 4 months and restricted activity since 1 month. There was history of fever accompanying the skin lesions. There was no similar history in the past and no history of contact with tuberculosis. There were no similar complaints in any other family members.

On examination, he was febrile, sick looking, had pallor with generalized lymphadenopathy. His vital parameters were within normal limits and anthropometry was normal. He had hepato-splenomegaly (liver 4cm below right costal margin with liver span of 9cm, spleen 2cm below left costal margin). Local examination showed multiple concentric, targetoid lesions with ring ulcerations, crusting surface of margins studded with vesicles, involving both thighs and buttocks **(*Fig.* 1).**

Investigations revealed a polymorphic leucocytosis (total count 27900 cells/mm^3; polymorphs 80%, lymphocytes 20%), microcytic hypochromic anemia (hemoglobin 7.3 g%), and thrombocytosis (platelets 720000 cells/mm^3). He had a raised ESR (60/110 mm) and a high C-reactive protein (168.8 mg/L). His renal and liver function tests were normal. Anti-nuclear antibody was negative. Mantoux test (10 TU) was negative and HIV Elisa was non-reactive. His immunoglobulin profile was also normal. Chest roentgenogram was normal and ultrasonogram of abdomen revealed mild hepatomegaly. His blood culture did not grow any organism. Bone marrow cytology revealed features of pure red cell aplasia (striking reduction in the erythroid progenitors with a few scattered



(*a*)

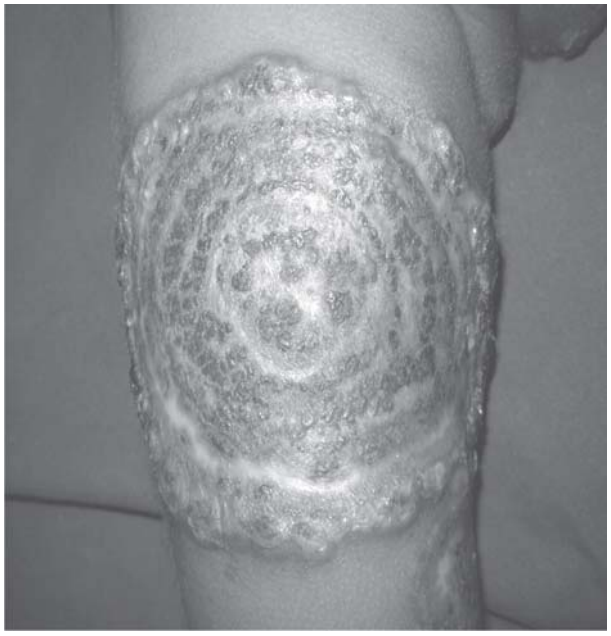


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FIG. 1 *Cutaneous lesions – multiple concentric, targetoid lesions showing extensive ulceration with a ragged undermined edge and crusting surface of margins studded with vesicles involving both thighs and buttocks.*

normoblast), and the skin biopsy of the lesion (***Fig.*2**) showed focal ulceration with sub-acute inflammation and superficial dermal abscess suggestive of superficial granulomatous pyoderma (abscess with sheets of neutrophils and karyorrhectic debris with few multinucleate giant cells and fibrinoid changes).

With the above clinical picture and laboratory findings, a final diagnosis of Pyoderma gangrenosum with pure red cell aplasia was made

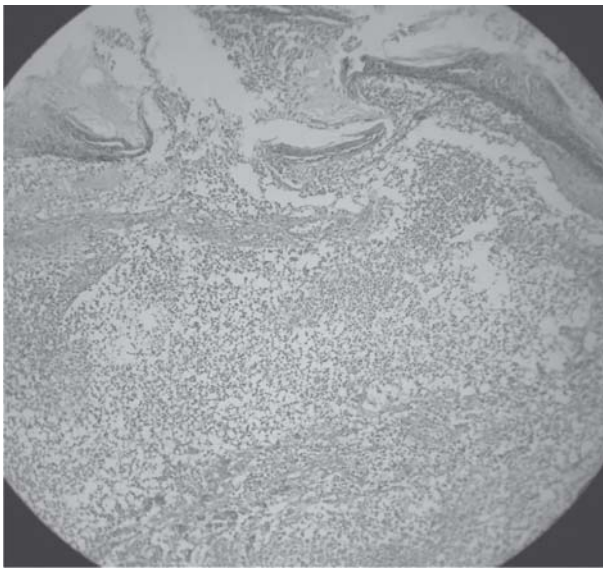

FIG. 2 *Skin lesion biopsy - 'superficial granulomatous pyoderma' – abscess with sheets of neutrophils & karyorrhectic debris with few multinucleate giant cells & fibrinoid changes.*

and the boy was treated with intravenous antibiotics (amoxyclav) for 1 week, and oral steroids (prednisolone 1mg/kg/day). He was also transfused with packed red blood cells and started on iron and folic acid supplementation. Fever settled in 4 days, with a dramatic healing of skin lesions in 10 days. He became ambulant and was discharged on oral prednisolone. Within a period of one month, the skin lesions had healed significantly and he had normal hemoglobin (11g%). His spleen was not palpable and the liver span had normalized. The steroids were tapered and stopped. On a four-month follow up, the child is doing well with no recurrences of the condition with normal hemoglobin levels.

DISCUSSION

Pyoderma gangrenosum (PG) is defined as a destructive necrotizing non-inflammatory ulceration of the skin clinically starting with sterile pustules that rapidly progress and turn into painful ulcers of variable depth and size with undermined violaceous borders(1). The disease is more common in adults aged between 30 to 50 years and only less than 4% of PG is seen in children(2,3).

Approximately 50-70% of PG is associated with an underlying systemic disease(4), and the asso-

ciated clinical conditions commonly include inflammatory bowel disease, rheumatoid arthritis, regional enteritis, chronic hepatitis, hematological disorders like myelogenous leukemia, acute lymphoblastic leukemia, multiple myeloma, polycythemia vera, myeloid metaplasia, pure red cell aplasia, and also diabetes mellitus and immunodeficiency(5). The lesions may precede, occur simultaneously or follow the above clinical conditions and healing of pyoderma gangrenosum lesions after successful control of the systemic disease have also been reported(6).

There are 4 major clinical classification types of PG: Ulcerative PG, Pustular PG, Bullous PG and the Vegetative PG(1). Our case had chronic non-painful ulceration, absent violaceous border with slow evolution - a form of superficial PG confined to the skin and known as 'superficial granulomatous pyoderma'- a vegetative type of PG which is usually not associated with systemic disease.

The diagnosis of PG is one of exclusion(6). The management of this disorder begins with treatment of any underlying disease and local or systemic glucocorticoids or immunomodulating therapies(1,6). Management of skin lesions include (*a*) local wound care, (*b*) topical therapy - topical steroids, cromolyn sodium, and nicotine, and (*c*) systemic therapy - systemic steroids, azathioprine, cyclosporine, tacrolimus, mycophenolate mofetil, methotrexate, chlorambucil, thalidomide, colchicines, cyclophosphamide, dapsone, minocycline, sulfapyridine, and TNF-μ inhibitors. Pulse methylprednisolone, pulse cyclophosphamide, and intravenous immunoglobulin have also shown benefit. Surgical treatment is useful only in extreme conditions(1,6,7).

Our child presented with progressive skin lesions, fever and pallor, and had no symptoms and signs of arthritis, gastrointestinal disease and malignancy. The diagnosis was made based on the skin biopsy and bone marrow cytology. The relation between pyoderma gangrenosum and pure red cell aplasia is found to be rare in the literature. Only two adult cases (a 72-year old man and an 80-year old woman) have been reported so far with this rare association(8,9) and no cases reported in the pediatric age group. This type of pure red cell aplasia is often associated with neoplasia, mostly thymoma. It has been reported earlier only in adults. The presence of serum inhibitors to erythropoiesis and the response to steroid therapy suggests an autoimmune phenomenon.

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