Mutilating pyoderma gangrenosum in an infant: A case report

Mary Rose Lim-del Mindo MD, DPDS; Daisy King-Ismael MD, FPDS

Pyoderma gangrenosum (PG) is a rare, inflammatory, ulcerative skin condition characterized by an ulcer with advancing violaceous border, negative cultures, unresponsiveness to antibiotics, the phenomenon of pathergy and histopathology showing neutrophilic infiltration. It usually occurs in all age groups but is rare among infants and very young children. A 6-month-old boy presented with papules which evolved into necrotic ulcers on the ears, perioral area, nose, neck, inguinal, scrotal, intergluteal and genital areas. The destruction of the ear cartilage made the clinical picture unusual and to the best of our knowledge, has rarely been reported as a feature of pyoderma gangrenosum in published case reports. The patient responded well to Hydrocortisone IV and tapering doses of oral prednisone. The long term effects in patients who developed infantile PG are still unknown and further follow-up of these unusual cases is recommended.

Keywords: Pyoderma Gangrenosum, neutrophilic dermatosis, pediatric dermatology

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare, inflammatory, ulcerative skin condition of unknown etiology. It has been reported that around 4% of the cases occur in infants and children. The diagnosis of PG is established by the clinical presentation and laboratory findings to exclude all alternative diagnoses. In children, the most common sites of involvement are the head, face, buttocks, genital and perianal area. The most common type of infantile PG reported to date is the ulcerative type. Ulcerative PG evolves as an inflammatory pustule that rapidly ulcerates. It can occur at sites of trivial trauma such as needlestick, inoculation site and insect bites (the pathergy phenomenon). The lesions progress rapidly into a marginalized and undermined ulcer with distinct borders and violaceous edge. It can also be excruciatingly painful. Typically, a thin atrophic, cribriform scar is left after healing. Atypical presentations, such as genital involvement, have been described previously in case reports but to our knowledge, the destruction of cartilage in cases of pyoderma gangrenosum has not yet been reported.

CASE REPORT

A previously well 6-month-old boy developed several erythematous papules on the left ear with no other accompanying symptoms. The lesions rapidly evolved into ulcers, which healed with scars. However, new papules appeared on the right ear, neck and nose despite oral antibiotics. Serum zinc levels were low and he was also given oral zinc. The chest x-ray showed parastrachal densities and patient was started on triple anti-Koch’s. Four months later, similar papules appeared on the nose and penis. These, later on, became necrotic ulcers and healed with the destruction of the nasal cartilage (Figure 1a) and disappearance of the penile shaft (Figure 1b). CBC showed leukocytosis and thrombocytosis. The culture of the oral mucosa revealed heavy growth of Enterobacter cloacae while the culture of the perianal area revealed heavy growth of Klebsiella oxytoca. Both specimens were negative for fungi or acid-fast bacilli. He was given topical and oral antibiotics. The ulcers improved and resolved with hyperpigmentation and atrophic scarring.

At 1 year of age, he developed fever and cough. There was again the appearance of new lesions despite the continuous treatment of zinc, oral and topical antibiotics and anti-Koch’s regimen. Chest x-ray showed pneumonia so he was given IV antibiotics. Chest CT scan revealed thickened tracheal wall with suspicious bronchial wall thickening and segmental atelectasis of the right lobe. Rapid plasma reagin (RPR) test was non-reactive but treponemal antibody (FTA-ABS) test was positive. He was given Penicillin G (54,000 IU mkd) for 10 days. New lesions were noted to appear on sites where the diaper was tight. A skin biopsy taken from the erythematous border of an ulcer on the sacral area showed focal loss of epidermis and dermal necrosis, moderate superficial to deep dermal perivascular and periadnexal mixed cell infiltrate consisting of lymphocytes, histiocytes, neutrophils, and few eosinophils (Figure 2a-b). Histopathological assessment was “consistent with pyoderma gangrenosum.” Prednisone (0.6 mkd) was started and improvement of the ulcers was noted. Two weeks later, he developed a productive cough with difficulty of breathing and was subsequently admitted. There were new ulcers and erosions noted over the arms, ears and neck. He was intubated and was started on IV antibiotics and hydrocortisone (40 mkd). Chest x-ray revealed pneumonia. The patient had anemia and blood transfusions were done. The previous areas of trauma including the sites of intravenous line on the scalp (Figure 1c), endotracheal tube insertion and the neck, and over areas where adhesives were applied developed lesions. The lesions later on dried up and IV hydrocortisone was shifted to tapering doses of oral prednisone. The lesions completely resolved. The patient was discharged.
DISCUSSION

Pyoderma gangrenosum is protean in its clinical expression, with variable clinical presentation according to type and stages of the disease.\(^3\) The four morphological variants of PG are ulcerative, pustular, bullous, and vegetative. As mentioned earlier, the most common type of infantile PG is ulcerative. The buttocks or perineum are commonly involved.\(^2\) Pyoderma gangrenosum in childhood demonstrate certain characteristic features. In children, the ulcers usually begin as pustules.\(^4\) In our patient, the lesions started as papules. In children, the lesions are also seen on the head and face, buttocks, perianal and genital regions.\(^4\) Infants tend to have perianal or genital involvement.\(^5\) In literature, the involvement of cartilage was never reported. In our patient, the major lesions of PG occurred on the ears, nose, mouth, perioral area, neck, inguinal, intergluteal and genital areas. The phenomenon of pathergy (the development of a new inflammatory lesion at the site of trauma) is described in 25% of patients with ulcerative pyoderma gangrenosum.\(^6\) In our patient, new ulcers appeared over sites of the intravenous line, endotracheal tube insertion and over areas where adhesives were applied. The key point in making a diagnosis of PG is the exclusion of other causes of cutaneous ulcers through biopsy, culture, and clinical acumen.\(^7\) The lesions in our patient presented with sharply marginated, undermined border.

Initially, our patient had been treated as a case of acrodermatitis enteropathica due to the low zinc levels. However, lesions progressed despite prolonged zinc supplement. In acrodermatitis enteropathica, zinc replacement therapy results in rapid clinical improvement and takes only a few days to a week to detect a significant change in serum zinc levels.\(^8\) The patient also completed the standard 6 months regimen of anti-Koch’s and was given several IV and oral antibiotics, however, the patient still had recurrent cough and colds. He had a positive treponemal test and a negative nontreponemal test. However, a single positive treponemal test cannot be solely relied upon as a means of diagnosing syphilis. The characteristic lesions of syphilis should also be
present. In a study by Peter and colleagues, their findings illustrated that false-positive or borderline FTA-ABS results can also be associated with a wide variety of viral infections including adenoviruses, coxsackievirus B, cytomegalovirus, herpes simplex virus (genital infection), influenza, mumps, parainfluenza, rubella, and rubella.⁹,¹⁰ Our patient presented with a persistent cough and colds unresponsive to anti-Koch’s and antibiotics. The positive treponemal test could be a false positive result.

The histopathological findings of pyoderma gangrenosum are non-specific. Biopsies taken from the central portion of the lesion show a central necrotizing suppurrative inflammation, usually with ulceration and those taken from the peripheral areas show a peripheral lymphocytic vascular reaction comprising perivascular and intramural lymphocytic infiltrates, usually without fibrin deposition or mural necrosis. Tissue neutrophilia with epithelial undermining and ulceration in the absence of leukocytoclastic vasculitis, fungal, bacterial or mycobacterial organisms highly implicates pyoderma gangrenosum.¹² Histopathologic findings in our patient from a specimen taken from the erythematous border of an ulcer on the sacral area, revealed focal loss of epidermis and dermal necrosis and with higher magnification showed moderate superficial to deep dermal perivascular and periannexal mixed cell infiltrate consisting of lymphocytes, histiocytes, neutrophils and few eosinophils.

PG is associated with systemic illnesses in about 50% of the adult cases, however, newborns rarely present with systemic diseases. Involvement of the lungs is the most common cutaneous manifestation of PG. It is characterized by patchy infiltrates or interstitial pneumonitis.¹² Our patient had episodes of cough and colds with interstitial infiltrates seen radiographically that was diagnosed to be tuberculosis and pneumonia.

Since PG is a rare condition, most treatment recommendations are based on experience. These often include combinations of oral corticosteroids and other medications that can inhibit the immune system. Systemic steroid therapy (1-2 mg/kg per day) remains the treatment of choice in most patients.⁶,¹² Corticosteroids remain the cornerstone of management in pediatric PG. Our infant’s new lesions responded well to prednisone.

REFERENCES
