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Pyoderma Gangrenosum Initially Presenting as an Ulceration of the Ear Lobule

Pyoderma Gangrenosum (PG) was first described in 1916 as “*phagedenisme geometrique*”, after a French dermatologist observed rapidly progressing, cutaneous necrotic lesions with sharp borders.¹ In 1930, Brunsting and his colleagues at the Mayo Clinic coined the term Pyoderma Gangrenosum, because it was initially thought to arise from staphylococcal and streptococcal infections which were observed in 5 of their patients.² The exact etiology and pathogenesis is still unknown. To date, only a few cases of PG have been shown to affect the ears, all showing no gender or age predilection.³ We report another such case.

CASE REPORT

A three-year-old girl presented at the emergency room with a non-healing, erythematous papule over her left ear lobule, allegedly following an ear piercing one month prior. She was initially treated at another institution with oral antibiotics. Despite treatment, her mother noted rapid worsening of the lesion, eventually developing into a painful ulceration and affecting the left eyelid as well. At the time of examination, the patient presented with a painful, necrotic plaque around the left eyelid with serpiginous borders (*Figure 1*) and ear lobule with erythematous, advancing borders (*Figure 2A, B*). There were no systemic co-morbidities noted. The working diagnosis then was necrotizing fasciitis and she was immediately started on systemic intravenous antibiotics which she did not respond to. Laboratory tests showed elevated CRP, but procalcitonin, C-ANCA and ANA were all normal. Tissue cultures of both eyelid and earlobe, as well as blood cultures, revealed no growth. Wedge biopsy of the eyelid ulceration revealed neutrophilic dermatitis. Biopsy of the ear lobule revealed suppurative granulomatous dermatitis with secondary leucocytoclastic vasculitis. Further workups for infection and possible systemic diseases were all unremarkable. A pathology test was negative. A diagnosis of pyoderma gangrenosum was made after excluding systemic and infectious causes. The patient was started on systemic prednisone at a dose of 1mg/kg/day which she slowly responded to. Surgical reconstruction of the earlobe was to be planned once the ulceration completely healed; unfortunately, this patient was lost to follow-up.

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Figure 1. Ulcerative lesions around an erythematous margin and serpiginous borders involving the upper eyelid



Figure 2. Rapidly progressive ulceration with areas of necrosis around the left **A.** preauricular; and **B.** post auricular area.

DISCUSSION

Pyoderma gangrenosum is rare in childhood, with approximately 4% of PG cases seen in infants and children usually affecting the head and perineal area.⁴ Studies have also shown that there is a female preponderance because reactive neutrophilic dermatoses are known to affect females.⁵ The disease arising from a neutrophilic process is the well-accepted etiology. With advances in biologic therapies, it has been found that it may be related to overexpression of growth factors IL-8, IL-18, IL-16, and TNF- α . Metabolic oscillations and aberrant neutrophil trafficking is also observed.⁶ Clinically, it begins as a painful nodule that may be seen after episodes of trauma. Pain is a prominent feature in the pediatric population. In the classic lesion, the pustule progresses to a necrotic ulceration with irregular, red-purplish inflammatory borders and a purulent or bloody exudate.⁷ They may present anywhere in the body, usually in multiples sites, as compared to the adult population where lesions are solitary and observed in the extremities.⁸ Aside from the classic presentation, other PG subtypes are pustular, bullous, vegetative, peristomal, genital, infantile and extracutaneous.⁹

In most cases, a biopsy is warranted to rule out other causes for the cutaneous lesion. Specimens can also be sent for bacterial and fungal cultures. Around 20% of patients present with pathergy, where inciting trauma (such as a biopsy or venipuncture) forms a new lesion.¹⁰ The histopathologic findings seen in the classic lesion is ulcerative associated with dense neutrophilic infiltrates. Su *et al.*¹⁰ proposed a diagnostic criterion for PG, where the major criteria is a rapidly enlarging, painful ulceration in the absence of any other cause

for the lesion. Since then, modifications to this criterion have been made; however, PG remains to be a diagnosis of exclusion.¹⁰ The key to diagnosis is a thorough history with emphasis on a history of pathergy, associated pain, and the presence of associated systemic diseases. When present in the head and neck, differential diagnoses such as infected preauricular cysts or sinuses, dissecting folliculitis, ulcerating basal cell carcinoma, and trigeminal trophic syndrome should be considered.¹¹ PG often occurs in isolation but may be associated with other systemic conditions in up to 50% of cases.³ In pediatric patients, it is most commonly associated with Inflammatory Bowel Disease (IBD), warranting investigation of gastrointestinal symptoms and further examination through endoscopy.⁸ It is also associated with hematologic disorders, such as myelodysplastic syndromes, leukemias and lymphoma.⁸ In the pre-adolescent population, PG is often idiopathic or associated with hematologic disorders, while IBD and PAPA (Pyogenic Arthritis, Pyoderma gangrenosum and Acne) are seen in the adolescent population.⁸ In the Philippines, one of the cases of PG published in literature is that of an adult female suffering from acute myelogenous leukemia who presented with the bullous type of the disease.¹¹ PG may also be the initial presentation of vasculitis such as Behcet disease and Takayasu's arteritis, rheumatoid arthritis, and neoplasms.¹²

Due to the rarity of the disease, there is no definite effective therapy for PG, and treatment has mainly been empiric. Treatment goals include relieving pain, controlling the inflammatory process and managing the underlying disease.⁹ Oral corticosteroids with a usual dose of 0.5-1 mg/



kg/day have been widely used in the pediatric and adult population due to the rapid response to treatment.⁹ Other reported systemic treatments are immunosuppressive drugs, thalidomide, minocycline, sulphasalazine, and dapsone. Infliximab and other biologic treatments have also been reported to provide clinical improvement after one infusion.⁸ Local wound care and pain control are also necessary.

In summary, pediatric pyoderma gangrenosum may present in multiple sites, and in atypical areas such as the head and neck. As a diagnosis of exclusion, a thorough workup is imperative to diagnose other etiologies and evaluate co-morbidities associated with the disease. Once diagnosed, aggressive immunosuppression should be started to facilitate complete recovery.

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