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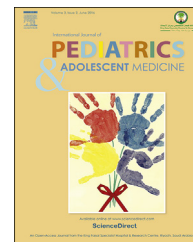


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INSTRUCTIVE CASE

Pyoderma gangrenosum with splenic involvement



Shaikha Jameel AlDossary^{a,b,*}, Tariq S. AlFawaz^b,
Awatif K. AlMutairi^c

^a Dammam University, Department of Pediatrics, King Fahd University Hospital, Khobar, Saudi Arabia

^b King Fahad Medical City, Riyadh, Saudi Arabia

^c Children Hospital King Fahad Medical City, Riyadh, Saudi Arabia

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KEYWORDS

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Abstract Pyoderma gangrenosum (PG) is a non-infectious disease characterized by necrotizing, ulcerative, and painful skin. The incidence of PG is approximately 3–10 cases per million people per year. Pyoderma gangrenosum is rarely observed in children, which accounts for less than 4%, but it primarily affects adults between the ages of 25 and 54 years old without gender preference. Here, we present a case report of a 16-month-old toddler diagnosed with pyoderma gangrenosum with splenic involvement. The young age of the patient and extracutaneous manifestation are of high interest.

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1. Introduction

Pyoderma gangrenosum (PG) is a non-infectious disease characterized by necrotizing, ulcerative, and painful skin

[1–3]. The incidence of PG is approximately 3–10 cases per million people per year [4]. Pyoderma gangrenosum is rarely observed in children, accounting for less than 4% of cases, but it primarily affects adults between the ages of 25 and 54 years old without gender preference. [1,2,5,6] However, the cause of pyoderma gangrenosum is poorly understood. Some researchers attributed the cause to dysfunction in the immune system [7,8]. In most cases, PG manifests as small lesions that become larger and spread. Pyoderma gangrenosum can be subcategorized as ulcerative, bullous, pustular, and vegetative [9,10]. A PG diagnosis is usually of exclusion, as there are no specific criteria to confirm diagnosis. Using systemic steroids is the most effective treatment of PG in children [1,2].

* Corresponding author. Dammam University, Department of Pediatrics, King Fahd University Hospital, Khobar, Saudi Arabia. Tel.: +966 503891101.

E-mail addresses: sh.al-dossary@windowslive.com (S.J. AlDossary), talfawaz@kfmc.med.sa (T.S. AlFawaz), akalmutairi@kfmc.med.sa (A.K. AlMutairi).

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Clinical information was retrieved via a retrospective review of medical records to determine clinical manifestation.

2. Case

A 16-month-old girl with no previous medical history presented with a fever associated with recurrent feet and hand abscesses that rapidly ruptured and formed deep ulcers. Skin examination at the ulcerous stage revealed multiple large tender necrotic oval ulcers with a purulent base and sharply defined violaceous undermined edges in the hands and feet (Fig. 1). Initial work-up showed leukocytosis with predominant neutrophils, an ESR of 83 mm/h and a CRP of 29.7 mg/L. Abscesses did not respond to antimicrobial therapy. Abdominal US revealed splenic abscess measuring 10.47 × 10.82 cm (Fig. 2). The culture from both skin lesions and the splenic lesion were negative, and the histopathology demonstrated characteristics of PG. However, all immune tests were within normal ranges. The patient was started on oral prednisolone (2 mg/kg/day) for two weeks and showed a good response, in which the fever resolved and all of the lesions healed.

3. Discussion

Pyoderma gangrenosum is neutrophilic dermatosis that is destructive in nature with unknown causes. More than half of patients with PG develop the disorder in association with an underlying systemic disease [11]. However, the diagnosis is made after excluding other causes of ulcerative skin lesions via histopathological findings. There is no diagnostic laboratory test that is specific for PG. The presence of neutrophils and increased ESR was observed in most of the affected patients [12]. In our patient, we made a diagnosis of pyoderma gangrenosum based on skin histopathology. However, idiopathic pyoderma gangrenosum was suggested due to exclusion of other usual associations.

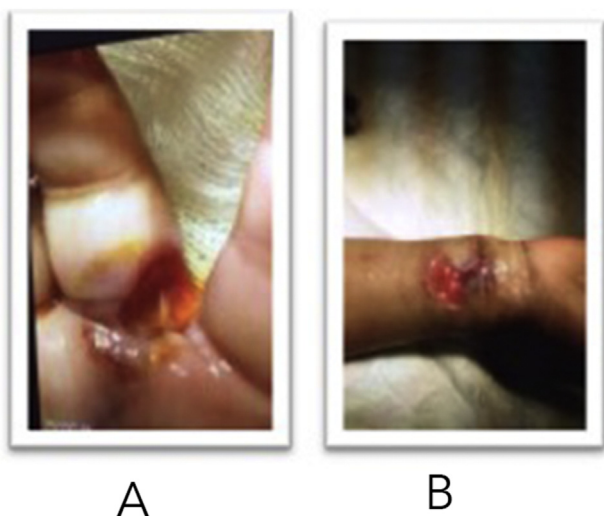


Figure 1 Abscess after healing forming an ulcer.

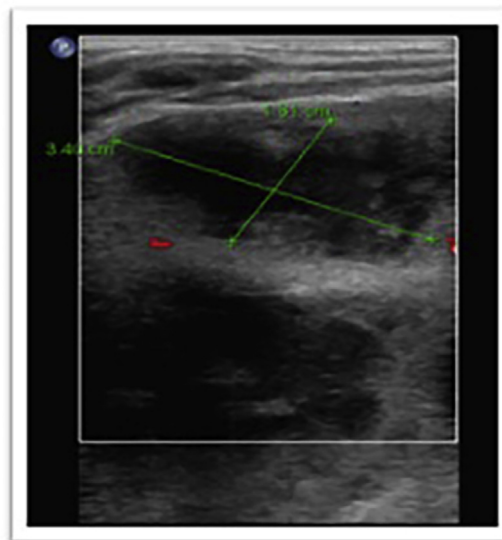


Figure 2 US abdomen showing splenic abscess.

Extracutaneous manifestation of PG in our patient was in the form of splenic involvement. Allen CP et al reported a child to have pyoderma gangrenosum with splenic and lung involvement [13], and Johnson JL et al reported a child with only splenic involvement [14]. There is limited data regarding extracutaneous manifestation of PG in the pediatric age group compared to adult patients, who frequently report this comorbidity.

Patients with superficial pyoderma gangrenosum are primarily treated with topical immunosuppressants (e.g., corticosteroids, calcineurin inhibitor, et al) whereas systemic treatment requires systemic therapy such as corticosteroids and cyclosporine [15]. Alternative therapies include dapsone, sulfapyridine, methotrexate, clofazimine, minocycline, colchicine, intravenous immunoglobulins, and anti-TNF agents [1]. This was described well in small uncontrolled studies and clinical experiences. Our patient improved completely after administration of systemic corticosteroid treatment, as observed by the rapid healing of the ulcer, resolving of the fever, and disappearance of the splenic abscess.

The number of pediatric patients with extracutaneous manifestation of PG is limited. Further studies and reports will enrich our knowledge in the clinical presentation of this disease as well as guidelines for improved management.

Conflict of interest

None to be declared by the authors.

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