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A typical Case of Pyoderma Gangrenous Revealing Bechet's Disease in An Infant

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Abstract

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis that presents with rapidly developing, painful skin ulcers hallmarked by undermined borders and peripheral erythema [1]. PG is often associated with a variety of other immune-mediated diseases, most commonly inflammatory bowel disease and rheumatoid arthritis. The cause of PG is not well understood, but PG is generally considered an autoinflammatory disorder.

We report the case of a 2-year-old infant with an atypical presentation of severe pyoderma gangrenous revealing behcet's disease.

PG is difficult to diagnose as several differential diagnoses are possible; in addition to clinical examination, laboratory tests of biopsied wound tissue are required for an accurate diagnosis, and new validated diagnostic criteria will facilitate the process.

Treatment of PG typically starts with fast-acting immunosuppressive drugs (corticosteroids and/or cyclosporine) to reduce inflammation followed by the addition of more slowly acting immunosuppressive drugs with superior adverse event profiles, including biologics (in particular, anti-tumour necrosis factor (TNF) agents. Appropriate wound care is also essential. Future research should focus on PG-specific outcome measures and PG quality-of-life studies.

Observation

24-month-old girl with good psychomotor development with a mother followed for Behçet's disease under treatment and the notion of four episodes of recurrent oral canker sores in infants.

She was brought by her parents for an extensive skin ulceration in the buttock (posterior side of the thigh) evolving for 8 months in the form of a confluent pustular rash with raised and extensive edges developing under a fever of 39 ° C, with notions of a single episode of chronic diarrhea following a dietary error (introduction of goat's milk at the age of 3 months); the lesions were rapidly extensive, resistant to local treatment and several systemic antibiotics. On clinical examination, we noted the atypical presence of multiple ulcerations (figure 1) in the same place (the left buttock) of various sizes ranging from 3 to 12 cm along the major axis, with raised border and fibrinous base (figure 1), these ulcers were located in the left buttock with magma of ipsilateral inguinal lenticular lymphadenopathy.

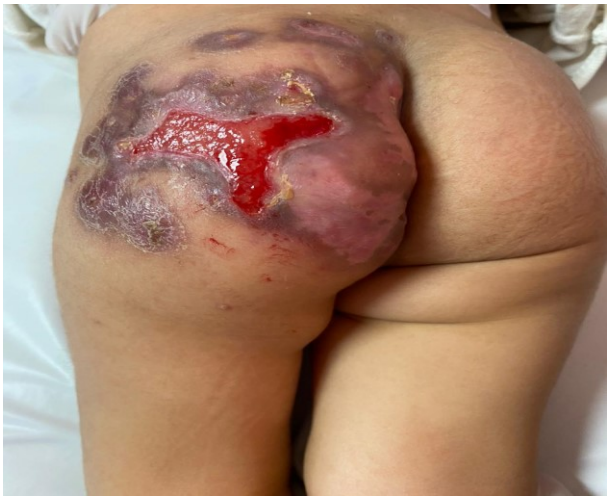

Figure 1 : Ulcerated lesion, with a hemorrhagic base, and excavated irregular borders, on the buttock.

A pathergic test was negative when a blood sample was taken from the right hand. The rest of the somatic examination (especially abdominal and joint) was normal.

The anatomopathological study revealed an acanthotic epidermis on the edges of the ulceration along with the presence of a pyogenic granuloma strongly infiltrated by neutrophils at the base of the ulceration (Fig. 3). The smear hemogram revealed microcytic hypochromic anemia with normal ferritinemia at 19; the sedimentation rate was 40 mm during the first hour and the C-reactive protein level was 54 mg / L. The following examinations were normal: the myelogram not revealing any abnormalities, electrophoresis of the proteins, detection of antinuclear antibodies, anti-neutrophil cyto-plasm (ANCA), anti-Saccha-romyces cerevisiae (ASCA) and factor rheumatoid, total immunoglobulin (Ig) E assay, celiac disease serologies: AC antitransglutaminases type IGA, total IGA, AC antiendomysium, human immunodeficiency virus (HIV) serology, abdominal ultrasound, eso-gastro-duo fibroscopy denale, colonoscopy determination of lymphocyte subpopulations, quantiferon TBK, BK sputum and TBK xpert gene research on skin biopsy as well as research for atypical mycobacteria on skin biopsy. TBK converting enzyme and quantifieron came back negative. Research for HLAB51 came back positive.

The chest x-ray had demonstrated an interstitial syndrome supplemented by a thoracic CT which revealed intraparenchymtous micro-nodules under pleural and centrilobular with a scattered bronchiolar arrangement in the two pulmonary fields.

This presentation made it possible to retain the diagnosis of Bechet disease revealed by pyoderma ganrenausum. Systemic corticosteroid therapy (1 mg / kg / day) allowed complete recovery over a period of 3 months with the lesions gradually reaping after the 5th month of treatment. An immunosuppressant (mycophenolate mofetil 600 mg / m2 / day) was administered to the patient and good compliance with vaccination according to the national vaccination program and with good clinical and biological progress and subsequent degression of corticosteroid therapy (Figure 4).

The originality of our observation lies in the rare association of PG with BM and the occurrence of their ulcerations in the same place when it comes to children. Behcet's disease was strongly suspected due to the repeated oral aphthosis, was more likely to have positive HLAB51 antigen, and was retained in our patient according to the criteria of the International Study Group for Behcet 's disease. This little girl presented one major criterion and two minor criteria (oral aphthosis and pseudofolliculitis); as well as the suggestive clinical appearance and the absence of any other infectious or vascular cause. Skin histology was therefore helpful, showing the presence of a dermal infiltrate rich in neutrophils (Figure 3).

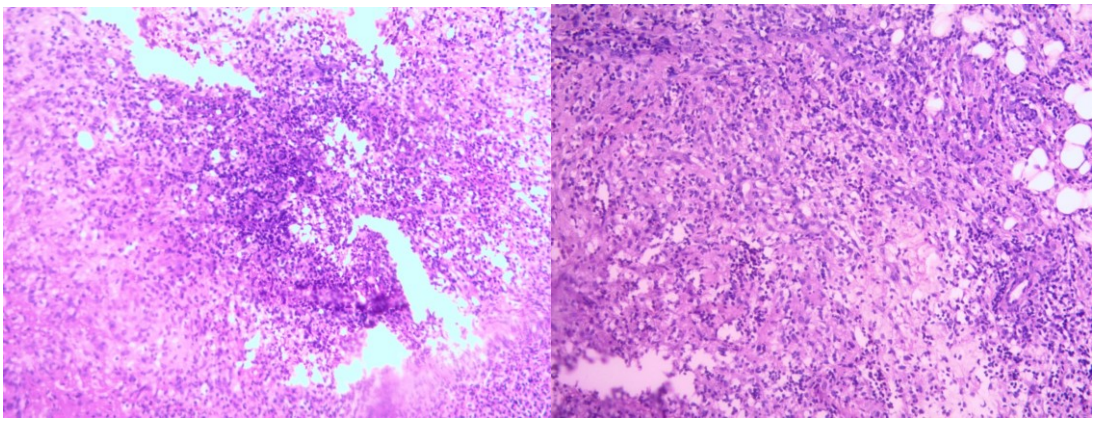

Figure 3: microscopic images at high magnification HE staining, dermal and hypodermic infiltration by an intense inflammatory infiltrate rich in altered polynuclear neutrophils producing in places images of micro abscess in favor of pyoderma gangrenosum, with no signs of malignancy.

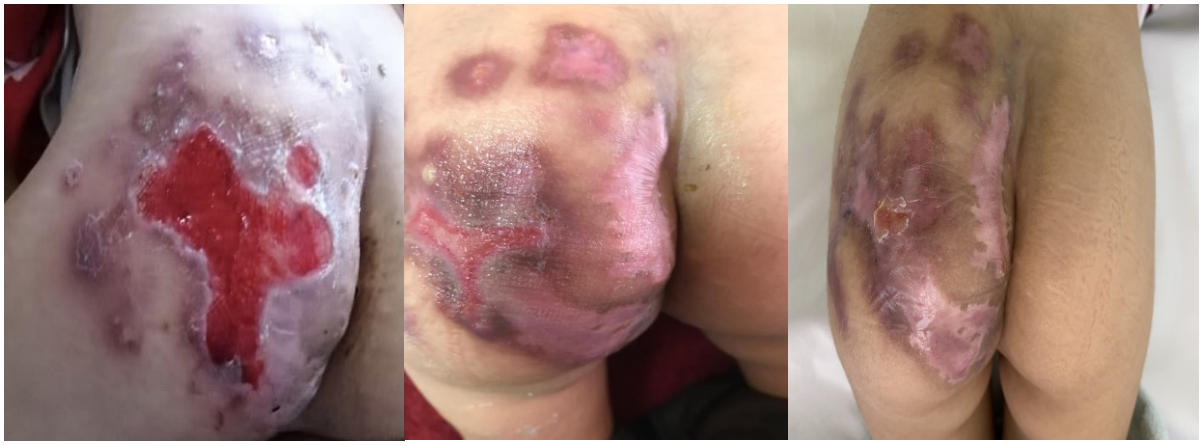

Figure 4: Clinical course of lesions after initiation of treatment.

Discussion

PG is a rare neutrophilic dermatosis impacting children. In a series of 180 cases identified at the Mayo Clinic, only 8 cases (4%) were under 15 years old [1]. Localization to the lower extremities is a common feature between children and adulhow. However, Graham et al. [2] reported other preferred locations when it comes to children, especially the head (26.1%), buttocks (15%), the perianal and genital area (7%). The pathergic test is a common feature in almost half of the cases (during vaccination, sampling, debridement) [3]. In our observation, the occurrence of ulcerations was atypical. Furthermore, it remained in the same site accompanied with an association of mouth ulcers, but whithout any ophthalmologic involvement. However, no article in literature has reported this association. The diagnosis of PG is essentially clinical (Table I) [4], no histological lesion is pathognomonic, yet, skin biopsy seems necessary in order to rule out other causes of skin ulcerations [5]. Su et al. [6] proposed diagnostic criteria (Table II) of which 2 major criteria and at least 2 minor criteria make it possible to retain this diagnosis. In literature, several diseases can proceed, accompany or follow the course of PG, mainly ulcerative colitis. In Graham's and al's series [2,14], 26.6% of children diagnosed with PG had no associated pathology.

Table 1: Clinico --- histological characteristics of pyoderma gangrenosum [4].

Clinical characteristics	Histological characteristics
Recurrent painful lesions (pustules, bubbles, ulcerations) arthralgia, myalgia, Positive pathergic test Ulcerations with active border under mined	Neutrophilic infiltrates

Table 2: Proposed diagnostic criteria for ulcerative pyoderma gangrenosum in its classic form [6].

Major criteria	Minor criteria
Rapid development of a painful skin ulceration with an irregular purplish border, under mined and necrotic; exclusion of other causes of skin ulceration	History suggestive of pathergia or clinical appearance of riddled scars Presence of associated systemic disease Anatomopathological data Response to systemic corticosteroids

In its typical form, the PG achieves a superficial ulceration of purple edges, arciform, raised, detached with several locasitions at the level of the body. Paradoxically, there is neither lymphadenopathy nor lymphangitis; the pain is variable [11,12]. In cases of corticosteroid-resistant PGs, the use of other immunosuppressants such as tacrolimus, ciclosporin and mycophenolate mofetil may be effective. Currently, anti-TNFa are a promising treatment for refractory PG [16]. The prognosis for PG depends on the associated disease and the severity of the clinical form of PG. Regarding BM, its occurrence among children is associated with early and severe visceral involvement [5,10].

Several publications have classified PG among systemic autoinflammatory diseases, secondary to a systemic effect of certain pro-inflammatory cytokines (IL-1b, IL8, IL6) in innate immune cells, in particular polynuclear neutrophils

[4,7]. Hence the need for systemic treatment. The best treatment is based on corticosteroid therapy (1 to 2 mg / kg / day) with a slowly decreasing regime. The combination of corticosteroids and ciclosporin has also been described [8]. In cases of corticosteroid-resistant childhood PG, the use of treatment alternatives may be effective, including dapsone, sulfapyridine, methotrexate, clofazimine, minocycline, and colchicine. Currently, the use of an anti-TNFa (tumor necrosis factor alpha), supported by the pathogenesis of auto-inflammation, is a promising treatment for refractory PG [9,13].

Conclusion

Through this observation, we describe an atypique case of PG, among an infant, revealing Bechet's disease and we underline the diagnostic difficulty when faced with a case of a child with persistent skin ulcers, which can delay the diagnosis and involve the functional or vital prognosis.

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