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Abstract

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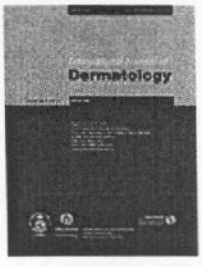

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Bullous pyoderma gangrenosum: a presentation of childhood Behcet's disease

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Bullous pyoderma gangrenosum: a presentation of childhood Behcet's disease

An 8-year-old girl presented with a history of recurrent painful skin lesions of 7 years' duration. The skin lesions started in the form of tender papules and nodules, which evolved into clear, fluid-filled, flaccid vesicles and bullae and ruptured spontaneously to leave behind a raw, ulcerated area. A history of recurrent oral and genital ulceration was present. The patient had been on oral steroids for about 5 years.

Examination revealed an underweight and malnourished child with cushingoid features. Multiple, asymmetric, extremely tender, flaccid bullae and well-demarcated ulcers with necrotic slough and hyperpigmented, erythematous, irregular margins were seen distributed over the body, more commonly over the trunk and proximal extremities (Fig. 1). The palms, soles, scalp, and face were spared. Post-inflammatory atrophic scars were also seen over the body. The oral and genital mucosa showed multiple, well-defined, shallow ulcers with an erythematous hue and slough-covered base. Nikolsky's sign was negative and no purpuric lesions were seen. A pathergy test was positive. Eye examination showed bilateral, mild-exposure keratitis. The patient was uncooperative for slit lamp and fundus examination.

On investigation, the hemoglobin was 9.6 g/dL, the total leukocyte count was 144×10^3/L, and the differential count showed 98% neutrophils with coarse granules. The erythrocyte sedimentation rate was 40 mm/h. Peripheral blood smear revealed normocytic, normochromic anemia with neutrophilic leukocytosis. The leukocyte alkaline phosphatase score was 367, suggesting a leukemoid reaction (normal, 22–155). The urine showed traces of albumin. The C-reactive protein level was 96 mg/L (normal, 0–6 mg/L). Rheumatoid factor was 32 IU/mL (normal, 0–10 IU/mL). Bulla fluid cytology revealed mainly neutrophils and degenerated cells. Blood and pus culture showed no growth of organisms. Immunoglobulin (Ig) assay revealed the following: IgA, 171.9 mg/dL (normal, 33–202 mg/dL); IgG, 1645 mg/dL (normal, 635–1280 mg/dL); IgM, 259 mg/dL (normal,

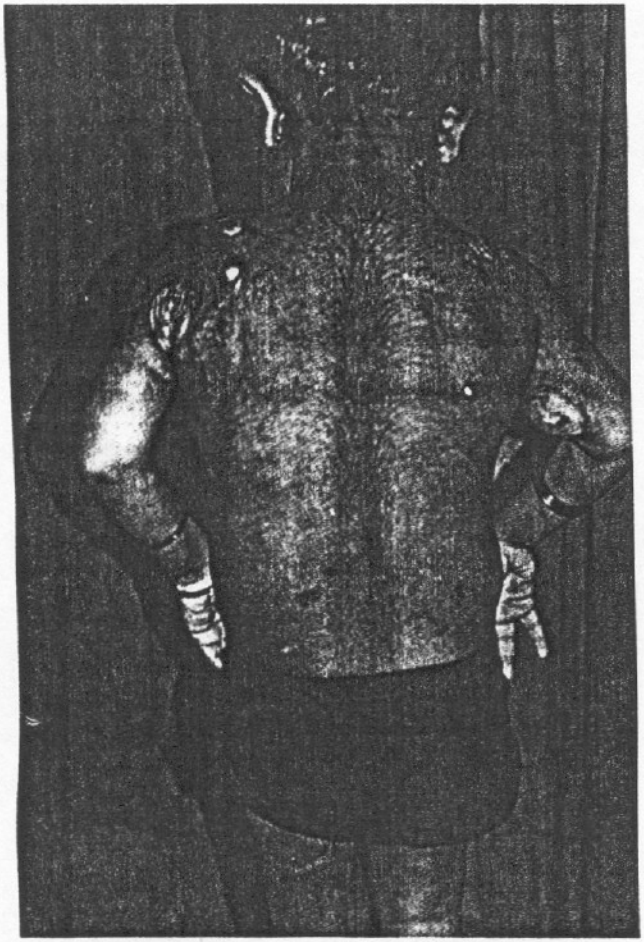

Figure 1 Multiple, asymmetric, extremely tender, flaccid bullae distributed over the trunk and proximal extremities

40–70 mg/dL). Bone marrow aspiration revealed myeloid precursor cells with coarse basophilic granules. Skin biopsy showed neutrophilic dermatosis with panniculitis. Chest X-ray and ultrasonography (USG) of the abdomen were normal.

Discussion

Behcet's disease is a multisystem inflammatory disorder of unknown origin, characterized by recurrent oral and genital ulcerations, ocular and cutaneous lesions, arthritis, and central nervous system and vascular disease. There is no pathognomonic laboratory test, but there are clinical criteria to assist in establishing the diagnosis.[1]

Recurrent oral and genital ulcers with bullous pyoderma gangrenosum and positive pathergy test established the rare diagnosis of Behcet's disease in our patient by fulfilling the O'Duffy criteria and International Study Group criteria for the diagnosis of Behcet's disease.[2]

Usually, the onset of Behcet's disease occurs between 20 and 30 years of age and is very uncommon in children.[3] The clinical spectrum of childhood Behcet's disease resembles that of adult disease; however, the disease seems to run a less severe course in children. Children with Behcet's disease have significantly less genital ulcers, less vascular thrombosis, and more non-specific gastrointestinal symptoms, as well as central nervous system involvement and arthralgia. A relatively high prevalence of uveitis is found in children.[4] Oral ulceration is the most frequent major sign and therefore should not be neglected as it may signal Behcet's disease.[1]

Various kinds of cutaneous lesion appear in patients with Behcet's disease: erythema nodosum-like lesions, papulopustular eruption, erythema multiforme-like lesions, thrombophlebitis, Sweet's syndrome, and pyoderma gangrenosum.[5] Bullous pyoderma gangrenosum, an atypical variant of pyoderma gangrenosum, is known to be associated with myelodysplastic syndromes.[6] Hitherto, to the best of our knowledge, bullous pyoderma gangrenosum has not been reported to be the presentation of childhood Behcet's disease.

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References

1 Lee LA. Behcet disease. *Semin Cutan Med Surg* 2001; 20: 53–57.
2 Ghate JV, Jorizzo JL. Behcet's disease and complex aphthosis. *J Am Acad Dermatol* 1999; 40: 1–13.
3 Kim DK, Chang SN, Bang D, *et al.* Clinical analysis of 40 cases of childhood onset Behcet's disease. *Pediatr Dermatol* 1994; 11: 95–101.
4 Krause I, Uziel Y, Guedj D, *et al.* Childhood Behcet's disease: clinical features and comparisons with adult onset disease. *Rheumatology (Oxford)* 1999; 38: 457–462.
5 Lee ES, Bang D, Lee S. Dermatological manifestation of Behcet's disease. *Yonsei Med J* 1997; 38: 380–389.
6 Suzuki Y, Kuroda K, Kojima T, *et al.* Unusual manifestation of myelodysplastic syndrome. *Br J Dermatol* 1995; 133: 483–486.