Pyoderma gangrenosum (PG) is a rare, inflammatory, neutrophilic disorder, first described in 1908 by Brocq (1). The pathogenesis of PG is not yet fully understood and, due to a lack of definitive diagnostic criteria, it is considered a diagnosis of exclusion (2). Suggested mechanisms leading to development of PG include neutrophilic dysfunction, genetic influence and a pathological inflammatory process (3–5). PG affects both men and women at all ages, with a prevalence peak between 50 and 60 years old (6). In up to 65% of cases of PG the disease is associated with an underlying systemic disorder (7). Treatment consists of wound care, pain control, and topical and systemic immunosuppressive therapy (2). We present here a rare case of PG in a splenectomy incision in a patient with haemolytic anaemia, which was treated successfully with systemic steroids. In addition, a literature review of similar cases is presented.

**CASE REPORT**

A 49-year-old woman, with no significant medical history, was admitted to the department of dermatology due to postoperative complications, which started 6 days after laparotomic splenectomy. Nine months before the surgery the patient was admitted to hospital due to paleness and excessive fatigue. On admission, full blood count revealed severe haemolytic anaemia with poikilocytosis, anisocytosis and spherocytosis. Serological analysis and further blood tests did not reveal any abnormalities. Bone marrow examination (myelogram and trephine biopsy) disclosed increased erythropoiesis with signs of dysplasia and excessive numbers of megakaryocytes. In addition, abdominal ultrasound revealed splenomegaly with numerous hyperechogenic areas and widened splenic vein. Spherocytosis was diagnosed based on the clinical picture and blood analysis. After 3 months and a few hospitalizations due to haemolytic crises, haematologists decided to remove the patient’s spleen. Difficulties in wound healing were observed shortly after the splenectomy. After unsuccessful therapy with empirical antibiotics, the woman was referred to the department of dermatology for consultation. On admission, 3 round ulcers with irregular, raised, purple, inflammatory borders and surrounding erythema were observed. Two of the ulcers were localized at the beginning and end of the surgical incision, and the third, similar ulcer, near the drainage (Fig. 1A). PG was diagnosed based on the clinical picture and anamnesis. The patient received 500 mg intravenous methylprednisolone daily for 5 consecutive days. Afterwards therapy was continued with oral prednisone, 40 mg/day. On the follow-up, after 4 weeks of therapy, the lesions healed completely (Fig. 1B).

**DISCUSSION**

PG is a rare neutrophilic dermatosis, in the course of which patients develop solitary or multiple painful skin ulceration with characteristic erythematous active border (8). The clinical presentation and course of PG often vary greatly between cases and, therefore, it is frequently misdiagnosed. The clinical variants of PG include ulcerative, bullous, pustular, vegetative, peristomal and surgical subtypes (2). The pathogenesis of PG is not fully understood; however, it is commonly associated with underlying systemic conditions. Authors often mention its co-occurrence with inflammatory bowel disease (ulcerous colitis and Crohn’s disease), rheumatological (polyarthritis), and haematological disorders (leukaemias, myelodysplastic syndromes) (2, 9, 10). Moreover, Al Ghazal et al. (11), in their paper indicated a necessity to look for additional, possible, unreported predisposing comorbidities, such as anaemia, obesity or diabetes. Furthermore, even less frequently, PG development with no predisposing disease or associated comorbidity is possible. The exaggeration of a skin injury, which may occur after minor trauma, termed pathergy phenomenon, may be seen in one-third of patients with PG. According to the study by Tolkachjov
et al. (12), the majority of patients (66%) who develop postoperative PG do not have any predisposing systemic condition. In those cases, ulceration at the incision site appears 6.86 ± 4.95 days after surgery (12). In the current patient the time until development of lesions was shorter and took 4 days. However, in the remainder of the cases reported, the time was 6–7 days. The lesions may affect almost every anatomical area: breasts, abdominal and chest wall, lower and upper extremities, with equal distribution among both sexes. Diagnosis of PG is difficult due to the lack of specific markers and histopathology. The optimal evaluation should involve the patient’s history, physical examination and skin biopsy (2). Diagnostic criteria, proposed by Mavarakis et al. (13), include 1 major criterion (neutrophilic infiltrate in skin biopsy) and 8 minor criteria (exclusion of infection; pathergy; history of inflammatory bowel disease or inflammatory arthritis; history of papule, pustule, or vesicle ulcerating within 4 days of appearing; peripheral erythema, undermining border, and tenderness at ulceration site; multiple ulcerations, at least one on an anterior lower leg; cribiform or “wrinkled paper” scarr(s) at healed ulcer sites; and decreased ulcer size within 1 month of initiating immunosuppressive medication(s)) (13). In the current case we decided not to take a biopsy, because the lesions had direct connection with a surgical injury. Since pathergy phenomenon was suspected, any additional trauma could cause further development of PG and deterioration of the patient’s health. Moreover, the diagnosis could be made on a clinical basis, due to a typical clinical picture, patient history of systemic disorder, and no response to antibiotic therapy. Treatment of PG presents a challenge for clinicians, as there are only 2 randomized controlled trials and no international guidelines. First-line therapy consists of analgesia, local wound care and topical treatments, which include corticosteroids or tacrolimus ointments. In more severe cases the patients are given oral steroids, cyclosporine, colchicine, sulphasalazine, dapsone or thalidomide. Lately, there is increasing evidence of the use of biological treatment in the management of PG. Among them, infliximab and adalimumab have shown the biggest efficacy in the wound healing, but empirical antibiotics, sometimes with negative swabs. It is important to emphasize, that a mistake in diagnosis may delay treatment and hence prolong patient’s symptoms.

The authors have no conflicts of interest to declare.

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