An Uncommon Case of the Acute Disseminated Haemorrhagic Bullous Type of Dermatitis Ulcerosa

Sir,

Dermatitis ulcerosa is an uncommon, destructive inflammatory and ulcerating skin disease of unknown cause. It is frequently the cutaneous manifestation of a systemic disease, such as ulcerative colitis, Crohn's disease, polyarthritis, gammopathy and some other conditions (1, 2). As the lesions of dermatitis ulcerosa do not exhibit a specific histopathology, and as other laboratory findings have also been variable and inconsistent, the clinical picture remains the cornerstone for diagnosis of this disease. An atypical bullous type of dermatitis ulcerosa has been recognized to herald preleukemic and leukemic states (3). It has an acute onset and is characterized by steadily enlarging, soft papules and blue-gray, haemorrhagic bullae. We report on a bullous type of ulcerative dermatitis in a patient with multiple systemic diseases, without evidence of leukemia, and an uncommon course of recovery.

CASE REPORT

The patient, a 38-year-old man with liver cirrhosis since 1989 and diabetes since 1995, was admitted to hospital because of an acute pyelonephritis caused by a concrement of the ureter in May 1996. The patient received antibiotics (ciprofloxacin, ofloxacin) and an ureteral pig-tail catheter for relief of the concrement. Due to a concomitant disorder of coagulation heparine, dexamethason and prednisolone were given. After dismissal he developed multiple painful papules and nodules on the buttocks and legs, rapidly leading to ulceration, fever and weariness. When admitted to our department, he was in a septic-like condition with fever up to 39°C. Mouth opening was severely reduced and showed a red, oedematous mucosa with an aphtha on the tip of the tongue. On the buttock, spreading out to the dorsal upper leg, were two 10×15 cm-sized, painful, ulcerating lesions with bullae on the undermined red margin, discharging haemorrhagic exsudate (Fig. 1). Around the rima ani two irregular necrotic demarcated ulcerations were localised. Two crusty reddish ulcerations were on the left knee and on the left lower leg. Furthermore both legs showed multiple solitary purple papules and haemorrhagic lesions. Some of them were spreading out and ulcerated quickly, discharging a serosanguinous exsudate. Blood examination, including p-ANCA and c-ANCA, was normal except for elevated BSR, white blood count (10.8/µl), C-reactive protein (107 mg/l) and thrombocytopenia (81,000/µl), anaemia, signs of hepatic damage, lowered calcium and phosphate, hypergammaglobulinaemia, hypalbuminaemia, reduced zinc and elevated cardiolipin-IgG. Urine analysis revealed massive erythrocyturia, proteinuria, albuminuria and elevated alpha-1-microglobulin. Blood culture, culture of blister fluids and Gram's stain were negative. ECG, echocardiography and X-ray of the chest were normal. Abdominal ultrasound and computer tomography of the chest and the abdomen revealed ascites, liver cirrhosis and splenomegaly. Bone marrow showed signs of asymptomatic thrombocytopenia of a megakaryocyte pattern. The histological examination proved oedema, massive neutrophilic inflammation, bullous haemorrhage and necrosis of epidermis and cutis. Furthermore, we saw engorgement and fibrinoid necrosis of small- and medium-sized vessels. Immunohistopathology did not reveal a specific pattern. As the findings of the mouth mucosa pointed towards a gingivostomatitis herpetica, acyclovir 3×500 mg was given i.v. For protection of bacterial superinfection and the known pyelonephritis cefotaxim 3×2 g and flucloxacillin 3×2 g i.v. were applied. Zinc deficiency was balanced. For prophylaxis of mycotic superinfection and therapy of gastrointestinal candidosis, fluconazol 200 mg daily was supplied. After 1 week of this therapy the fever disappeared and the patient's condition was stable. When the clinical aspects of dermatitis ulcerosa dominated, a therapy with sulfasalazine 500 mg orally was started and

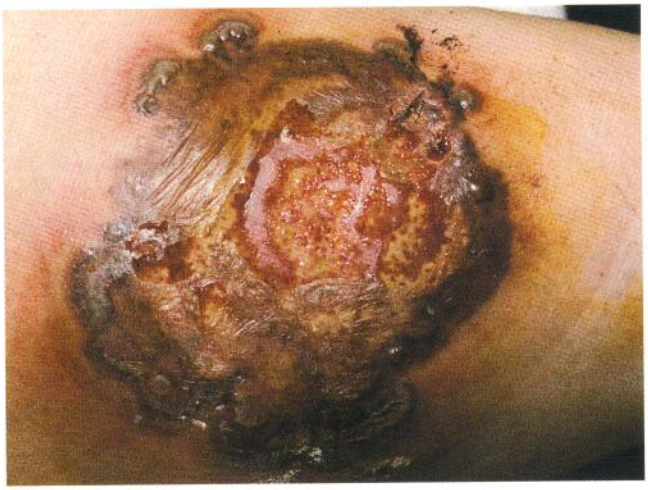

Fig. 1. Painful, bullous, ulcerating lesion on the dorsal upper leg, surrounded by an undermined red margin on admission.

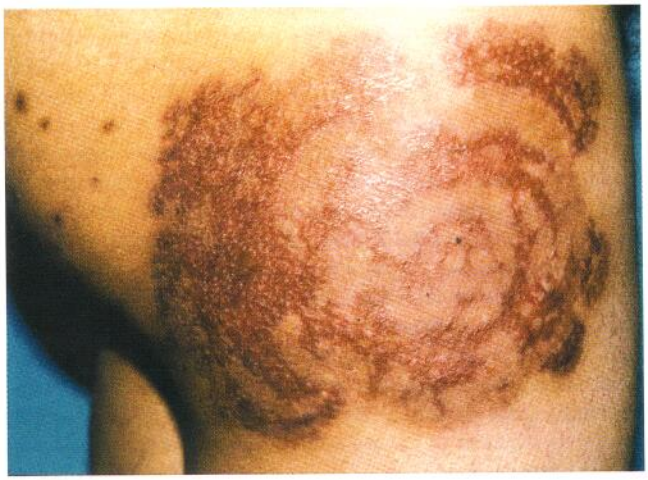

Fig. 2. Lesion of the upper leg 2 months after onset of the disease, with a ringworm-like epithelization.

increased to 2 × 500 mg daily. The ulcers epithelized in a ringworm-like pattern, and no new lesion appeared (Fig. 2). Unfortunately, the patient developed a toxic bone marrow hypoplasia, leading to pancytopenia, and the sulfasalazine had to be discontinued after 2 weeks. The healing continued by intensive antiseptic and granulation supporting therapy. The patient could be discharged in a stable condition without any lesions.

DISCUSSION

According to the clinical course differential diagnosis revealed necrotizing vasculitis of the lower plexus, metastatic calcinosis, streptococcal necrotizing fasciitis and large cell-Ki-1-anaplastic lymphoma. Diseases known so far to be associated with dermatitis ulcerosa could be excluded. In particular no leukemia or malignant lymphoma, often seen in the bullous type of dermatitis ulcerosa, could be diagnosed (3). One paper reports a 34-year-old woman with bullous dermatitis ulcerosa, who was free of symptoms for 7 months and developed acute myeloid leukemia accompanied by recurrence of the skin disease 12 months after diagnosis of dermatitis ulcerosa (4). This underlines the necessity of a long-term observation of the patient's condition, including blood count. Septic courses, anaemia and affection of bone marrow are usually considered to have an association with leukemia (3), but in our patient anaemia and bone marrow affection could also be result of liver cirrhosis. Abnormalities in cell-mediated immunity, humoral immunity and chemotactic functions may play a role (4) and are even more likely in a patient with liver cirrhosis. As the patient, when admitted to our hospital, was in a septic-like condition and had a history of pyelonephritis and disorder of coagulation, corticosteroids were not a therapeutical option (6). Because of the patient's multimorbidity and the improved cutaneous status we hesitated to start another systemic therapy after discontinuation of sulfasalazine and limited therapy to intensive local therapy. Whether the severe pyelonephritis and herpetic gingivostomatitis, and a severe liver cirrhosis as well, were the initiating cumulative factors of the severe course of dermatitis ulcerosa has to be discussed. It has also been observed in association with active, chronic hepatitis (7) and diabetes (8). The clinical course of recovery without relapse of lesions despite of discontinuation of sulfasalazine supports the probability of an infectious cause. There is only one author who also observed associated infectious diseases such as sinusitis, tonsillitis and amoebic dysentery with complete healing after therapy of the infectious disease (9). Whether the liver dysfunction or the presence of infectious foci was the ultimate cause may be revealed by the follow-up of the patient.

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