Acta Dermatovenerol Croat 2014;22(1):44-47 CASE REPORT

Extreme Dermatology – the Intensive Care Skills of Dermatologists in Three Case Presentations of Acute Skin Failure

Carmen Maria Sălăvăstru[1], Elena Severin[1], Mihaela Moisa[1], Klaus Fritz[2], George-Sorin Țiplica[1]

[1]2nd Clinic of Dermatology – Colentina Clinical Hospital, Bucharest, Romania; [2]Dermatology Clinic, Landau, Germany

Corresponding author:

Professor Carmen Maria Sălăvăstru, MD, PhD

2nd Clinic of Dermatology

Colentina Clinical Hospital

Bucharest

Romania

galati1968@yahoo.com

Received: May 8, 2013

Accepted: September 10, 2013

SUMMARY Dermatological emergencies include a number of clinical conditions usually accompanied by systemic symptoms that can lead to life-threatening complications.

From the broad spectrum of life-threatening dermatoses, three cases are presented: a case of febrile ulceronecrotic Mucha-Habermann disease (FUMHD), a case of pemphigus vulgaris mimicking Stevens-Johnson syndrome (SJS), and a case of toxic epidermal necrolysis (TEN). Those cases were considered extreme, and presented to illustrate the positive outcome of timely intensive dermatological care.

An interdisciplinary approach is essential in the diagnosis, treatment, management, and follow up of patients with life-threatening dermatoses.

KEY WORDS: life-threatening dermatoses, pemphigus vulgaris, toxic epidermal necrolysis, febrile ulceronecrotic Mucha-Habermann disease

INTRODUCTION

Dermatological emergencies include various generalized dermatoses that can cause disruption in the anatomy and physiology of the skin, leading to a number of complications known as "acute skin failure", a fatal syndrome which fulfills the definition of an emergency " - a risk perceived by a doctor or a patient to life, limb or the structure/function of an important organ of the body" (1,2). Prompt implementation of appropriate treatment for the various systemic complications of life-threatening dermatoses is mandatory. Between January 2011 and November 2012, 3,962 patients were admitted to the Clinic of which 129 (3.25%) required special therapeutic measures: 36 cases of pemphigus vulgaris (0.9%), 1 case of toxic epidermal necrolysis (0.02%), and 92 cases of angioedema (2.32%). The three most severe cases of life-threatening dermatoses were selected for presentation: a case of febrile ulceronecrotic Mucha-Habermann disease (FUMHD), a case of PV mimicking SJS and a case of toxic epidermal necrolysis (TEN), in order to raise the awareness of the complexity of treating such cases by clinical dermatologists.

CASE REPORTS

Case 1. A previously healthy 25-year-old male patient was admitted for a generalized eruption of erythemato-violaceous round-to-oval-shaped papular lesions, some of them coalescing in large polycyclic plaques. The disease had started 4 weeks earlier as erythematous macules, 10 days after the patient self-administrated tetracycline for a dental

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abscess. Routine laboratory investigations revealed leukocytosis, neutrophilia, lymphopenia, monocytosis, inflammatory syndrome, and highly increased levels of serum anti-hepatitis B (HB) antigen associated with low serum levels of hepatitis B virus (HBV) DNA. Bacteriological examination of the skin lesions indicated superinfection with *E. coli* and *Enterobacter spp*. The biopsy of a cutaneous lesion on the abdomen revealed spongiosis and parakeratosis of the epidermis with exocytosis of numerous lymphocytes and erythrocytes in the papillary dermis, associated with profound hematic and lympho-plasmocytic inflammatory infiltrate reaching the deeper portion of the dermis.

During the first 2 weeks of hospitalization, the patient developed daily febrile episodes in the 38.1-39.5°C range, accompanied by a local burning sensation. All blood cultures taken in the febrile period were sterile. After 7 days of hospitalization, the lesions became ulcero-necrotic and covered over 60% of the patient's body surface, primarily in the intertriginous and flexor areas, with sparing of the mucous membranes (Figure 1a). The diagnosis of FUMHD was established and systemic treatment was started with prednisone 0.5 mg/kg, erythromycin and 2 grams ceftazidime per day each, and analgesics and antipyretics. Topical treatment consisted of antiseptics, dermatocorticoids, antibiotics, and sterile greasy bandages for the intertriginous ulcerations. The patient was discharged from the hospital 6 weeks later: his cutaneous lesions gradually improved and slowly healed. At follow-up examinations performed after 1 and 6 months, the patient was in good general condition, but developed numerous hypertrophic and keloid scars (Figure 1b).

Case 2. A 43-year-old female patient was referred to our clinic for confluent erosive lesions covering 70% of the skin surface (Figure 2a), painful oral erosions, and impaired general condition, diagnosed as SJS. The patient denied having a history of significant diseases. The current disease had started 2 months ago, as painful erosions in the oral cavity with no improvement after a course of systemic antibiotics. One month after onset, a disseminated cutaneous eruption appeared, composed of flaccid blisters that gave rise to extensive denuded bleeding surfaces located especially on the back of the trunk, upper chest, and periorificial areas. The clinical suspicion of paraneoplastic pemphigus was excluded by CT imaging. Histopathology findings were suggestive of PV. Direct immunofluorescence demonstrated IgG and C3 positive staining in a network pattern (Figure 2b). Laboratory investigations showed anemia, hypoproteinaemia, severe hypoalbuminemia (the lowest value 1.1 g/dL; n.v.=3.4-4.8 g/dL), hypokalemia (the lowest value 1.85 mmol/L; n.v.=3.6-5.2 mmol/L), hypocalcemia, and inflammatory syndrome. Bacteriological examination of the skin lesions indicated superinfection with *E. coli* and *Pseudomonas aeruginosa*. One of the main therapeutic goals was the correction of fluid and electrolyte imbalances, but also treating the severe hypoalbuminemia. This was achieved by working closely with intensive care physicians. The treatment consisted of systemic corticosteroids (prednisone 2 mg/kg daily) with slow tapering of the dose. The introduction of azathioprine as a corticosteroid-sparing agent was not possible because the patient developed high fever. Broad spectrum antibiotics (ertapenem, teicoplanin, and colistin) were administered for the infection of the cutaneous lesions. Local

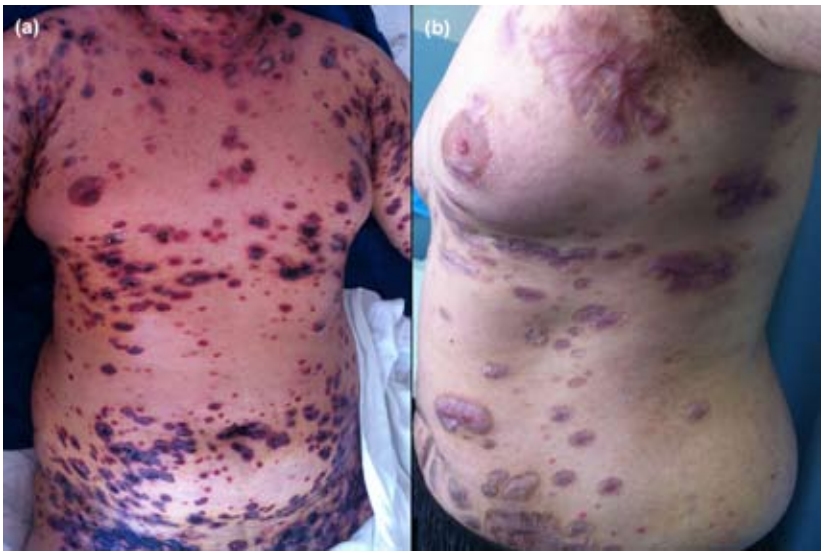

Figure 1. (a) Multiple prominent ulcero-necrotic lesions in the intertriginous areas. **(b)** Numerous hypertrophic and keloid scars at time of follow-up visit after 6 months.

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treatment consisted of antiseptics, antibiotics, and dermatocorticoids. Disease course during hospitalization was slowly favorable. The patient was discharged after 2 months of treatment, with cutaneous lesions having been almost completely reepithelized. At a follow-up visit after 1 year, the patient was in good general condition, with discrete hyperpigmented macular lesions on the trunk.

Case 3. A 52-year-old patient was referred to our clinic for a cutaneous eruption consisting of blisters and erosions covered by hematic crusts, distributed over the entire surface of the skin (Fig 3). He also had mucosal involvement of the palate, gingiva, tongue, and genital area. There was conjunctival hyperemia, abundant conjunctival secretion, photophobia, and ocular foreign body sensation. The eruption started on the face 3 days prior to admission. The patient had been diagnosed with bipolar affective disorder 2 years ago, and had been treated with lorazepam, valproic acid, olanzapine, and lamotrigine. The last two drugs were introduced two weeks before the onset of the eruption. Considering the sudden onset, disease course, fever, drug intake, and the body surface involved (over 60%), the diagnosis of TEN was established. The general treatment measures included: drugs discontinuation, blister puncture, removal of secretions, and daily local dressings. The treatment consisted of systemic corticotherapy and antibiotherapy, electrolytes replacement, parenteral nutrition, antihistamines, topical treatment with antiseptic solutions, and antibiotics. We also used artificial tears and eyewashes with antibiotics to treat corneal damage. After 3 weeks of hospitalization, the denuded areas where replaced by postlesional hyperpigmentations and few hematic crusts. During hospitalization, the patient had an unsuccessful suicide attempt.

DISCUSSION

FUMHD is a rare and sometimes lethal form (mortality 25%) of pityriasis lichenoides and varioliformis acuta, characterized by the dominance of necrotic skin lesions and high fever as well as other systemic symptoms, such as abdominal pain, arthritis, pulmonary, and central nervous system involvement (3,4). A recent review described 41 cases of FUMHD treated with different therapeutic agents (5). While the etiology of the disease remains unknown, one of the hypothesis proposes the idea that FUMHD is a hypersensitivity reaction after an infection (4,5). Problems encountered during patient hospitalization were: daily febrile episodes accompanied by severe burning sensation in skin lesions during the first two weeks, slowly improved by antipyretic treatment, large areas of ulceration in the folds at high risk of infection, and the association with newly diagnosed hepatitis B, a possible trigger for the disease. Under the course of systemic corticosteroids, erythromycin, and a careful local therapy, disease evolution was favorable. The patient became afebrile after the first two weeks, and was discharged after two months with cutaneous lesions partially *reepithelialized.*

PV can be a life-threatening autoimmune blistering disorder, with a mortality of 5-15%, in which extensive skin and mucosal lesions result in the failure of main skin functions, in particular the defense against infections (6). The major cause of death in these patients are severe infections, including septicemia, pneumonia, and cardiovascular disorder. The aim of the treatment is to stop the production of auto antibodies by administering corticosteroids, immunosuppressants, and, more recently, biologics such as rituximab (7).

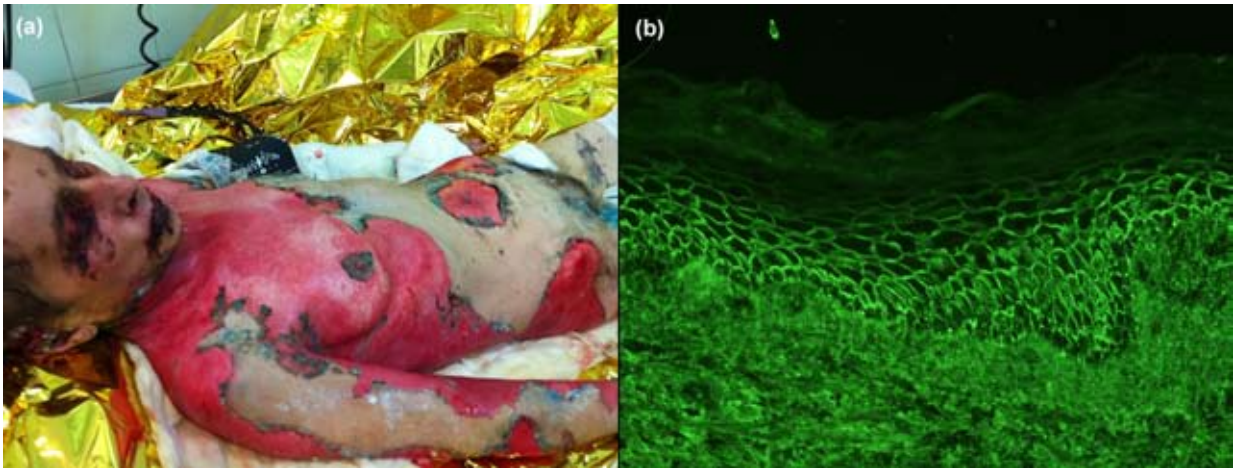

Figure 2. (a) Large erosive areas in a patient with PV. **(b)** Direct immunofluorescence positive for IgG in a network pattern.

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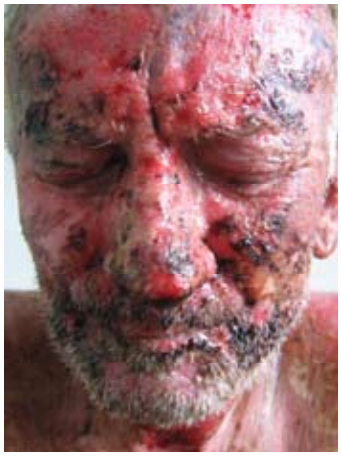

Figure 3. Erosions covered by hematic crusts on the entire surface of the face.

TEN is a life-threatening disease with sudden onset presenting as large epidermal denudation and mucosal involvement (8). The trigger is almost always a drug, and the affected cutaneous area is greater than 30%. Lamotrigine is a high risk drug in terms of TEN onset. The time between initial drug intake and the lesions onset is between 1 and 3 weeks (9). TEN can produce a great number of complications even when adequate treatment is implemented, and mortality may reach 30-40% (10). The patient presented in case 3 developed corneal damage with a risk of blinding. When TEN occurs due to psychiatric medication, the patient must be closely supervised and the causative medication discontinued. Systemic steroids can also induce psychiatric symptoms even at low doses (11).

For the presented cases, treatment priority was the correction and maintenance of haemodynamic equilibrium and electrolite balance by fluid and electrolyte replacement. Hypoalbuminemia caused by extensive areas of denuded skin was also addressed by creating a positive balance of nitrogen by enteral proteic nutrition and by treating the inflammatory syndrome. In order to compensate for the hypercatabolic state and to promote tissue healing, massive nutritional support was provided. With regard to sepsis, the suprainfection of the skin lesions imposed the introduction of broad spectrum antibiotics.

CONCLUSION

Life threatening dermatoses can frequent occur in a dermatologists' office. Many cases present immediate type reactions (eg. anaphylaxis, angioedema) or severe and acute bacterial, viral, fungal, or parasitic infections. In some severe cases, as those described in this paper, well-equipped units must provide optimal conditions for supportive care (fluid, electrolyte, and nutritional replacement), allowing patients to receive complex medical care tailored to disease severity.

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