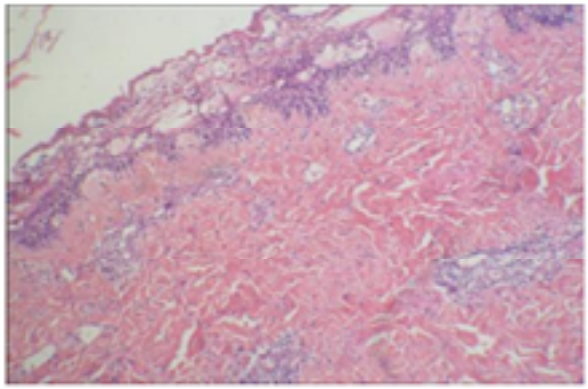
*Case Report*

Karelidze T., Katsitadze A.

# FEBRILE ULCERONECROTIC MUCHAHABERMANN DISEASE

TSMU, DEPARTMENT OF DERMATOLOGYAND VENEREOLOGY

Abstract We describe the case of a 64-years-old woman who was admitted for erysipelas of the face. Despite treatment, evolution was marked by the appearance of a necrotising ulcerative area in the centre of the erysipelas associated with local oedema and headache. A skin biopsy revealed a pityriasislichenoidesetvarioliformisacuta. Corticosteroids led to a rapid stabilisation of lesions, and after 6 months the patient shows only a small area of frontal hypopigmentation. The aetiology remains uncertain. There is no established standard treatment. We would like to draw attention of the medical and surgical specialists to this rare disease. Febrile UlceronecroticMucha-Habermann disease (FUMHD), first defined by Degos et al. in 1966, is a severe variant of pityriasislichenoides et varioliformisacuta (PLEVA). Characteristics of FUMHD include a fulminant course of the disease, painful ulceronecrotic erosions, fever and severe systemic manifestations. In current literature only 40 cases with an overall case fatality rate of 20% are described. Until today the knowledge about the etiology of this disease is only fragmentary, but an association with viral and lymphoproliferative diseases is being discussed. Case Report A 64-years-old woman is admitted for an infection in the forehead area. The infection began 10 days ago and the patient mentions a possible spider bite. At first, she applied a cream containing urea on the lesion. Her medical history contains no dermatological diseases, but she has suffered from Crohn's disease with no event for 14 years and without treatment. Her medication is composed of an antidepressant (mirtazapine) and a proton pump inhibitor (rabeprazole). At home, the initial treatment was composed of clindamycin (300 mg three times daily) and Fucicort cream (fusidic acid and betamethasone). The first clinical examination reveals an inflammatory wound with a diameter of 1 cm at the base of the scalp and frontal oedema extending to the eyes and nose. Her medical parameters were normal and laboratory tests revealed only a moderate inflammation (C reactive protein 15 mg/L normal value <5.0 mg/L). In front of the suspicion of erysipelas, antibiotic are continued intravenously. The evolution was marked on the day of admission by the rapid extension of the lesion and the appearance of necrosis . We noted the appearance of macula and papula away from the main lesion on the face and different parts of the body (back, abdomen and limbs). The palpation was painful but there was no sign of subcutaneous emphysema. The patient reported a headache, pruritus and a burning sensation. The general condition of the patient is preserved and its parameters are normal. She had no bowel disease symptoms. Given the lack of diagnosis and rapid progression of we performed a skin biopsy in different parts of the necrotic lesion and peripheral lesions. Histological examinations showed epidermal necrosis, the seat of many elements including inflam- matory mononuclear cells, many neutrophils, eosinophils and a few red blood cells. The superficial dermis shows a lichenoid infiltrate invading the basal layer which shows apoptotic and necrotic keratinocytes. There are also signs of leukocytoclasticvasculitis .The histopathological report concludes with a pityriasislichenoidesetvarioliformisacuta (PLEVA). The immunohistology found intradermal and intraepidermal lymphocytes CD8 and some macrophages. Treatment



Over the lack of response to treatment, on the day of the biopsy, systemic corticosteroids were started, intravenous methylprednisolone (120 mg) for 5 days relayed by oral form. The follow-up was marked by rapid stabilisation of the lesions after 1 week and local improvement after 1 month. Her home medical treatment consisted of a regression therapy of methylprednisolone (Medrol) and ointment including hydrocortisone butyrate (LocoidLipocream). Six months later, the patient presents only with a skin hypopigmentation area in the forehead . Although Mucha-Habermann disease is a severe variant of PLEVA, it differs by rapid and painful composed of macula, papula topped by scales. Thesepapula become vesicular-haemorrhagic pustules progressing to necrosis and ulceration. The ulceronecrotic form is characterised by rapid progression of necrotic papules that coalesce into large patches ulcerated. In general, lesions predominate on the trunk, roots and the ends of the limbs, but whole of the body can be affected including the mucous membranes. Systemic symptoms in the literature includes: high fever, neurological signs, asthenia, alteration of the clinical status, abdominal pain, diarrhoea, interstitial lung disease, cardiomyopathy, rheumatological manifestations, megaloblasticanaemiaand death. The causes of death are due to: pneumonia, pulmonary thromboembolism, cardiac arrest, sepsis, hypovolemic shock and massive thrombosis of the superior mesenteric artery.

**Keywords**: Febrile ulceronecroticMucha-Habermann disease; pityriasislichenoidesetvarioliformisacuta; fatal outcome; T cell monoclonality.

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*შემთხვევის აღწერა*

**კარელიძე თ., კაციტაძე ა.**
**მუხა-ჰაბერმანის ფებრილური წყლულოვან-ნეკროზული დაავადება**
**თსსუ, დერმატო - ვენეროლოგიის დეპარტამენტი**

მუხა-ჰაბერმანის ფებრილური წყლულოვან-ნეკროზული დაავადება წარმოადგენს მწვავე ლიქნოიდური და ვარი ოლი ფორმული პარაფსორიაზის რთულ ფორმას. დაავადება საკმაოდ იშვიათია და ხასიათდება კანის ნეკროზულ-წყლულოვანი დაზიანებებით, მაღალი ცხელებითა და სისტემური ცვლილებებით. მისი მკურნალობა საკმაოდ რთულია. ავტორების მიერ აღწერილია 64 წლის პაციენტის შემთხვევა, სადაც დიაგნოზი დაისვა ბიოფსიის შედეგის საფუძველზე და დაიწყო მკურნალობა კორტიკოსტეროიდით. მკურნალობა გაგრძელდა მედიკამენტის შემამცირებელი დოზითა და ადგილობრივი სტეროიდებით. ეს შემთხვევა ავღწერეთ, რადგან წარმოადგენს იშვიათ ფორმას, დამახასიათებელია მაღალი ცხელება,წყლულოვან-ნეკროზული ცვლილებები და, რაც მეტად საყურადღებოა, ხასიათდება ლეტალობით.