

Acquired Ulcero-Mutilating Bilateral Acro-Osteopathy (Bureau- Barrière Syndrome)

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Abstract

We present a 35-year-old male patient with Bureau-*Barrière* syndrome. Bureau-*Barrière* syndrome is an ulcero-mutilating acropathy almost invariably associated with excessive alcohol intake. It presents with a triad of trophic skin changes with recurrent ulcerations, bone lesions and nerve damage. The clinical presentation includes chronic painless plantar ulcerations with perilucerous hyperkeratosis, hyperhidrosis, livedoid skin colour, nail dystrophy, widening and infiltration of the toes and common interdigital mycoses. Other non-specific skin changes related to the alcohol consumption are commonly observed as well. The condition affects mainly middle-aged men suffering from alcoholism. Often a bilateral location at the lower limb of male alcoholics has been described, as in our patient. Successful treatment of the Bureau-*Barrière* syndrome requires an interdisciplinary approach. Cessation of alcohol intake and smoking is of paramount importance.

Citation: Tchernev G, Mangarov H, Lozev I, Pidakev I, Lotti T, Wollina U, Gianfaldoni S, Semkova K, Lotti J, França K, Batashki A. Acquired Ulcero-Mutilating Bilateral Acro-Osteopathy (Bureau-*Barrière* Syndrome). Open Access Maced J Med Sci. 2017 Jul 25; 5(4):558-560. <https://doi.org/10.3889/oamjms.2017.134>

Keywords: neuropathy; acquired; eczema; ulcerations; ulcero mutilating lesions; bilateral.

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Received: 17-Apr-2017; Revised: 23-Apr-2017; Accepted: 24-Apr-2017; Online first: 24-Jul-2017

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Funding: This research did not receive any financial support.

Competing Interests: The authors have declared that no competing interests exist.

We present a 35-year-old male patient, admitted urgently for the first time to the clinic of dermatology and dermatologic surgery with chronic deep ulceration on the toes of both feet, severe generalised itching, malaise, pain in the sacrum area and fever (Fig. 1a, 1b). The patient was hospitalised previously with the diagnosis of osteoarthritis purulenta digitorum pedis dextra, requiring surgical excision with synovectomy and sequesters removal. Also, his past medical history included childhood atopic dermatitis with allergic rhinoconjunctivitis. Examination revealed severe swelling of the toes of both feet with a deep ulceration to the depth of the

underlying tendon on the plantar surface of the left thumb (Fig. 1c). Superficial ulcerations were also observed on the plantar surface of the right thumb (Fig. 1d). The skin of both lower legs was markedly erythematous with an eczematous eruption, fine desquamation and yellowish secretion (Fig. 1b). The toenails showed bilateral onychodystrophic changes secondary to underlying onychomycosis without previous therapy (Fig. 1a, 1b). On the skin of the extensor surfaces of both hands, there were multiple excoriations, lichenification and solitary nummular plaques with a diameter of 1 to 3 cm. Alcohol-induced hepatic steatosis, chronic focal calculous cholecystitis

and splenomegaly were established by the laboratory screening. Significantly increased values of CRP (80.1 mg/l) and GGT (1969.0 IU/l) were observed. A considerably increased Anti-streptolysin titer from 1600 IU/ml (normal range up to 200 IU/ml) with fever suggested secondary erysipelas (cellulitis). Neurological clinical examination revealed polyneuropathy with distal hypoesthesia, impaired sense of space, decreased superficial and deep distal sensation and vegetative trophic changes. Electrophysiological studies showed evidence of axonal degeneration of motor fibres of n. peroneus, n. tibialis in the distal segments and sensory fibres of n. suralis, indicating severe distal sensorimotor polyneuropathy - axonal type. Radiculopathy of the sensory root S1 was established as an additional finding. Hemangiomas in L3 and L4, unrelated to the radicular symptoms were also observed.



Figure 1: Typical clinical findings in a patient with Bureau-Barrière syndrome

Despite the ulcerative changes in the thumb of the lower limbs, radiographic evidence of osteomyelitis was missing. Based on the medical history of osteoarthritis purulenta digitorum pedis, chronic bilateral ulceration of the toes with the initiation of unilateral elephantiasis of the thumb; vegetative disturbances (pronounced facial erythrosis and hyperhidrosis), history of regular alcohol consumption and bilateral symptomatic polyneuropathy with the acquired, non-familial form of ulcerative ulcero-mutilating bilateral acro-osteopathy type Bureau-Barrière. The therapy included Clindamycin 600 mg/3 x daily iv for seven days, followed by Ciprofloxacin 500 mg/2 x daily 5 days per os in combination with dual antihistamine therapy: levocetirizine, dihydrochloride 5 mg 1x/day, of the lower limbs (with metabolic - toxic genesis), the patient was diagnosed chloropyramine hydrochloride 20 mg 1x day intramuscularly; prednisolone 30 mg per day in a reduction regimen for 4 days; with topical application of iodine povidone 10% ointment for the ulcerations on both great toes; Clioquinolum/Flumethasonum cream for nummular eczematoid lesions 2x per day (Fig. 1d, 1f). A mechanical removal of the hyperkeratosis of both thumbs was also performed, after an initial good therapeutic response (Fig. 1d, 1e, 1f). The

polyneuropathy was treated with piracetam 1200 mg (1/1/0) for an initial period of 30 days in combination with pentoxifylline 400 mg (1/0/1) with subsequent dose reduction of the piracetam; Vitamin B12-1000 UI intramuscular application – 1x per day for 10 days, followed by 500 UI every 2 weeks for a total period of 3 months. A significant improvement of neurological symptoms was observed.

Bureau-Barrière syndrome is an ulcero-mutilating acropathy almost invariably associated with excessive alcohol intake. It presents with a triad of trophic skin changes with recurrent ulcerations, bone lesions and nerve damage. The clinical presentation includes chronic painless plantar ulcerations with periulcerous hyperkeratosis, hyperhidrosis, livedoid skin colour, nail dystrophy, widening and infiltration of the toes and common interdigital mycoses [1, 2]. Other non-specific skin changes related to the alcohol consumption are commonly observed as well. The condition affects mainly middle-aged men suffering from alcoholism [1, 3]. Often a bilateral location at the lower limb of male alcoholics has been described, as in our patient [2, 3].

Although the etiologic role of alcohol is well established, the exact pathogenesis is still unclear. The mechanical theory underlines microtraumatism as the main mechanism, whilst the sympathetic theory focuses on the vasomotility dysfunction. An integrative theory combines the former and the latter.

The differential diagnosis of Bureau-Barrière syndrome includes the inherited Thévenard's disease with a positive family history, Charcot's neuropathy in patients with diabetes mellitus, and syringomyelia. In the latter case, pain is lesser than joint of bone destruction suggest, that also means it is not completely painless [4]. Tethered cord syndrome is a rare condition with associated hyperhidrosis [5], and last not least infectious diseases such as borelliosis and leprosy have to be considered [6, 7].

Successful treatment of the Bureau-Barrière syndrome requires an interdisciplinary approach [1, 2]. Cessation of alcohol intake and smoking is of paramount importance.

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