

CORRESPONDENCE

Scleromyxedema diagnosis following unexplained encephalopathy

Scleromyxedema is a rare idiopathic disorder, characterized by papules in a thickened mucinous dermis, with fibroblastic proliferation and monoclonal paraproteinemia [1, 2]. Typically, scleromyxedema affects middle-aged men, has a chronic progressive course and frequently presents extracutaneous complications that generally occur after cutaneous disease [3].

A 67-year-old man, previously healthy, presented with vertigo, nausea and gait unsteadiness. Examination disclosed slow right phase rotatory nystagmus, without ataxia. Diagnosed with a positional benign vertigo, he was discharged with symptomatic medication. Three days later he suddenly lost consciousness from which he did not recover. At admission he was in a coma, with conjugate ocular movements and decerebrating posture with noxious stimuli, without fever or meningeal signs. MRI was normal and laboratory studies unremarkable. A few hours later he had a tonic-clonic generalized seizure. EEG showed slow and irregular background activity. CSF evidenced pleocytosis (1800 cells, 95% neutrophils), with normal protein and glucose contents, and negative microbiologic, virologic and serologic studies. He was prescribed valproate, cefotaxime, ampicillin, acyclovir and dexamethasone. He started to recover (d3), awake but with incomprehensible speech, very confused and agitated, without motor deficits; he had two more generalized seizures. LP was repeated (d7), and was completely normal. The patient slowly improved and, at day 11, he could speak and comprehend normally, but then started complex visual hallucinations: a two hectares field in the wall, smoke, and animals running. This improved spontaneously, with no complaints on dismissal (d14).

Six months later he presented a widespread symmetric eruption of 2-3 mm, firm, waxy, closely spaced papules predominating in the hands, forearms and forehead (*figure 1*). Physical examination also revealed diffuse erythema and induration of the skin, leading to decreased motility of the mouth and joints, especially the hands. Skin biopsy showed a diffuse mucin deposit in the dermis and marked fibroblastic proliferation. The most relevant results of the hematological investigation were IgG monoclonal gammopathy with λ light chains and absence of thyroid disease.

Several well-described CNS manifestations in scleromyxedema are encephalopathy, psychosis, stroke-like syndromes, seizures and, rarely, vertigo, memory loss, gait disturbance and dysarthria [3]. Also scleroderma-like lesions and Parkinson's disease have been described as possibly linked with exposure to pesticides [4]. There are reports of a specific "dermato-neuro" syndrome, in which fever, disturbed consciousness and seizures follow a flu-like illness [5]. CNS symptoms usually appear in patients

with previous cutaneous disease concurring with exacerbations of the later [3]. There are six reports describing neurological symptoms preceding cutaneous disease, in periods ranging from days till two years, with the following presentations: stroke-like syndrome, encephalopathy and encephalitis. In all patients neurological investigations were normal, except for EEG (slow diffuse background activity) and, occasionally, elevated CSF protein [3].

The mechanism by which the CNS is affected in scleromyxedema is unknown, but paraproteinemia and mucin cerebral deposition have been postulated [6]. These mechanisms, however, lack validity, some patients having no paraproteinemia with a negative brain biopsy [6]. An immunological abnormal response was recently documented, with increased interleukin 6 production and abnormal blood-brain barrier function [6].



Figure 1. Firm, waxy, closely spaced papules particularly visible in the forehead **A**), but also in the forearms **B**).

Our patient had a vertigo syndrome followed shortly by comatose state, seizures, CSF pleocytosis and, in the recovery phase, hallucinations. This case, on neurological grounds, remained a puzzle until the detection of cutaneous lesions. Although it is not possible to definitely link these two findings, previous reports and the absence of any other known neurological disorder which could account for such a presentation, after exhaustive investigation and proper follow-up, strongly suggest that they are caused by the same systemic condition, that is, scleromyxedema. After this, proper treatment and anticipation of other extracutaneous complications was undertaken. ■

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¹ Neurology Department,
Hospital de São Marcos, Braga, Portugal

² Dermatology Department,
Hospital de São Marcos, Braga, Portugal
alvmac@gmail.com

Margarida RODRIGUES¹
Álvaro MACHADO¹
Filipa VENTURA²
Maria Luz DUARTE²
Carla FERREIRA¹

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Linear atrophoderma of Moulin

A 42-year-old Caucasian woman presented a 5-year history of atrophic brown maculae on the left arm and trunk with an asymmetrical linear distribution forming an inverted U-shape from the breast area up the upper arm, an S-shape on the anterior abdomen, a V-shape on the back toward the median line (figure 1). The patient reported that the lesions developed few days after working under sun exposure in contact with parasiticide substances and tomato plants. There was no history of atopic dermatitis, psoriasis, or previous dermatitis. Past medical history was unremarkable. She denied any past treatment of the lesions. A biopsy revealed hyperpigmentation of epidermal basal cells, slight thickening of the collagen fibers in the mid-deep dermis with a rare perivascular lymphocytic infiltrate (figure 1). Laboratory findings were unremarkable except for an increase of hepatic enzymes compatible with hepatic steatosis. Antinuclear (ANA) and anti-DNA antibodies were negative and immunoglobulins were suggestive of previous common viral exanthematic diseases. The clinical picture, the histology, along with the lack of autoantibodies, were compatible with Linear Atrophoderma of Moulin (LAM) following Blaschko's lines (BL). Cytogenetic analysis found a normal karyotype without genetic mosaicisms. Following treatment with a high dose of vitamin E (400 UI/d) and topical clobetasol propionate, a slight improvement of the lesions was observed.



Figure 1. Asymmetrical S-shaped linear distribution on the left arm and trunk of the atrophic brown maculae. Histology of the skin biopsy characterized by the hyperpigmentation of epidermal basal cells and the slight thickening of the collagen fibers of the mid-deep dermis (H & E, $\times 150$).

Firstly described by Moulin *et al.* in 1992 in 5 patients [1], LAM is a distinct clinical entity characterized by acquired atrophic bandlike skin lesions that often show hyperpigmentation and always follow the lines of Blaschko. No preceding inflammation is noted, but a transient inflammatory stage is perhaps often unrecognized, and there is no induration or scleroderma. Usually the condition begins in childhood or adolescence, and there is no evidence of any long-term progression. Histopathologically, an irregular moderate hyperpigmentation of the lower part of the epidermis is found, along with a few perivascular lymphocytes in the dermis and slight thickening of the collagen bundles, as in our case [2]. The existence of LAM is controversial in its possible clinical overlap with linear scleroderma or morphea. Nevertheless, this latter is characterized by one or more linear streaks of progressive induration that can extend through the dermis, subcutaneous tissue, and muscle to the underlying bone, causing significant deformities [3]. The lack of autoantibodies, such as ANA, found in 73% of adult patients with linear scleroderma and the chronic and unvaried course make this diagnosis unlikely in our patient, leading to the more compatible diagnosis of LAM [4]. The cause and pathogenesis of this disorder remain