

# Lichen sclerosus et atrophicans, scleroderma *en coup de sabre* and Lyme borreliosis

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## Abstract

Lichen sclerosus et atrophicans (LSA) is a chronic, inflammatory skin disease of unknown etiology, characterized by atrophy.

We report a case of LSA with frontoparietal distribution, mimicking scleroderma *en coup de sabre*, causing scarring alopecia. The case was associated with *Borrelia* infection. The lesion improved with 2 cycles of antibiotic therapy with ceftriaxone 2 gr/day i.v for 21 days associated with UVA-1 therapy and local and systemic vitamin E supply (400 mg 2x/day *per os* for 3 months). This case stresses the importance of identifying clinical manifestations associated with Lyme disease and the use of tissue PCR to detect borrelial DNA in patients with these lesions, but characterized by negative serology for *Borrelia*.

## Introduction

Lichen sclerosus et atrophicans (LSA) is a chronic inflammatory muco-cutaneous disorder, characterized by sclerotic and atrophic lesions, most commonly found in adult women.<sup>1</sup>

It affects the anogenital area in 85-98% of the cases and less frequently the extragenital area.<sup>2</sup>

The most commonly affected extragenital areas are the neck and the shoulders,<sup>1</sup> but also the inner thighs, the submammary area, the wrist<sup>2</sup> and occasionally the oral mucosa.<sup>3</sup>

The involvement of the scalp is not frequent and its outcome could be similar to scarring alopecia, which could be the result of different diseases.<sup>4</sup>

The etiology of LSA is still unknown. Besides the genetic and local factors (Koebner phenomenon) and the autoimmune hypothesis, supported by the association of different autoimmune disorders, especially of thyroid origin (30% of cases),<sup>5,6</sup> an infectious hypothesis has also been proposed.<sup>7,8</sup>

## Case Report

A 57-year-old Caucasian woman presented with a history of asymptomatic frontoparietal lesion. Such lesion, which had been developing over the 3 previous years, was initially erythematous and became progressively atrophic and sclerodermic. The patient had been living in a highly endemic area for *Borrelia burgdorferi*, but she could not recall any tick bite or erythema chronicum migrans. She referred excellent health conditions. She also reported the simultaneous onset of migrating diffused myoarthralgias involving knees, hands, ankles, elbows, shoulders, as well as short-term memory impairment for two years, worsened previous seasonal insomnia for one year. Moreover, she presented migrating paresthesias involving the left side of the body for one month. The dermatological examination revealed an atrophic-sclerodermic lesion of about 10 cm in length and 3 cm in width, from the scalp to the centre of the forehead as reported in Figure 1. The skin was thin, inelastic, mother-of-pearl in shade, with erythematous margins of the lesion with subsequent scarring alopecia as well (Figure 1).

A skin biopsy was obtained from the scalp and after histological examination three different pathologists confirmed independently the diagnosis of LSA. It consisted of epidermal atrophy, oedema with superficial layer, hyperkeratosis and collagen production with cell rarefaction. The classical band of lymphocytes was present. Lymphoid infiltrate was observed even in the dermal-subepidermal junction indicating a possible later evolution of the LSA toward morphoea (Figure 2).

Serological IgM and IgG for *Borrelia burgdorferi* with enzyme-linked immunoassay test (ELISA) confirmed by Western Blot analysis was negative. Anti-nuclear antibodies (ANA), extractable nuclear antigen antibodies (ENA), anti-native DNA antibodies (n-DNA), anti-neutrophil cytoplasmic antibodies (ANCA) and erythrocyte sedimentation rate were negative.

PCR analysis for the detection of *Borrelia burgdorferi* was performed on DNA obtained from formalin-fixed paraffin-embedded skin biopsy, blood and urine as previously reported.<sup>9</sup> PCR analysis was positive in all the specimens. After 6 months the PCR was repeated on DNA from blood and resulted negative.

The diagnosis of Lyme borreliosis was made on the basis of clinical data, supported by the PCR positivity for *Borrelia* genome.

The patient underwent antibiotic treatment with 2 cycles ceftriaxone 2 gr/day i.v for 21 days. After about 6 months since the beginning of antibiotic therapy, the lesion had not progressed, all other clinical symptoms improved and blood PCR resulted negative. The patient

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received vitamin E 400 mg 2x/day *per os* for 3 months and applied topic vitamin E prior to UVA-1 therapy. UVA-1 treatment improved considerably her clinical condition. Thanks to this combined therapy a remarkable regression of the atrophic-sclerodermic lesion was observed.

## Discussion

This report describes a case of LSA, which was unusual for the involvement of the scalp. The lesion was mimicking scleroderma *en coup de sabre*, which is a frontal or frontoparietal linear morphoea characterized by a linear band of depressed atrophy on skin and scalp.



Figure 1. Lichen sclerosus et atrophicans with frontoparietal distribution, mimicking scleroderma *en coup de sabre*.

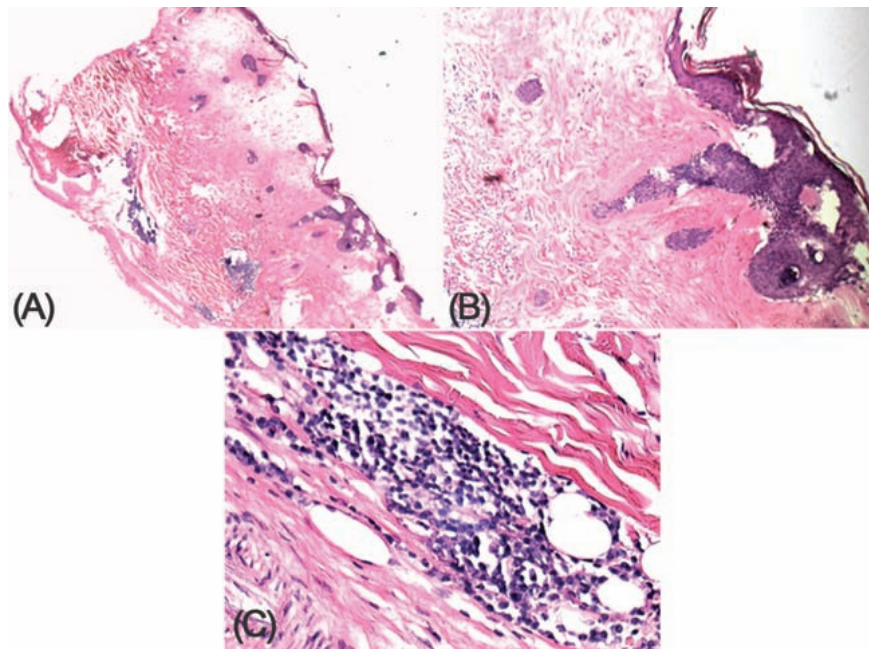


Figure 2. H&E stained section of the lesion (A) 2,5 x magnification of the entire histological section, (B) 20x magnification of the lichen sclerosus et atrophicus features (C) 40x magnification of the lymphatic infiltrate.

This lesion can frequently result in scarring alopecia.

The classical features of LSA<sup>10</sup> are represented by hypopigmented papules that coalesce into white plaques with epidermal atrophy. Although anogenital LSA is associated with a risk of 4-5% of squamous cell carcinoma, extragenital lesions do not seem to carry any risk of malignant degeneration.<sup>2</sup> The isolated linear frontoparietal involvement was described in few cases and may clinically simulate scleroderma *en coup de sabre*, requiring careful histopathological recognition.<sup>11</sup> In this case, LSA has been diagnosed independently by three different expert pathologists. It has already been reported that overlap of histologic features between LSA and morphea may occur, however in the reported case the clinical and the histologic features were not ambiguous of LSA. The possible later evolution toward morphea in this case, due to the migration of the lymphatic infiltrate, is not unusual since morphea and LSA may be closely related such that the latter could be considered the superficial expression of the same disease process which results in morphea.<sup>12</sup>

Regarding the possibility of an infectious etiology of LSA, since the first proposal by Aberer and Stanek in 1987<sup>7</sup> several European studies have debated on the association with *Borrelia* infection<sup>13</sup> with conflicting results. Some atrophic skin diseases have been proposed as manifestation of Lyme borreliosis with contradictory results.<sup>8,14</sup> The detection methods, the examined specimens, such as

sera, skin biopsies and urine, together with the different geographic region could explain the conflicting results on the association of *Borrelia* with morphea and lichen sclerosus et atrophicus.<sup>8</sup> Moreover in the manifestation of long standing infection of *Borrelia* the paucity of microorganisms could lead to a low detection rate by PCR, especially when the analysis is performed on archival biopsies.<sup>8</sup> No decision can be made to date as to whether *Bb* plays a role as causative agent of different types of circumscribed scleroderma and LSA. With regard to the disparate findings in different geographic areas, it can be speculated that LSA may be caused in some cases by *Bb* genotypes which are present in that area only.<sup>15</sup> To support this infectious etiology in endemic regions, several cases respond to antibiotic therapy for borreliosis, such as penicillin and ceftriaxone.<sup>16</sup>

The fact that in this case *Borrelia* DNA was detected both on DNA obtained from the biopsy and from blood and urine, strongly supports the hypothesis that *Borrelia* has a causative role on the onset of this unusual LSA. Moreover, the geographical location, Middle Europe, has been highly associated with *Borrelia* prevalence.<sup>8</sup> Indeed, it is well known that there are significant geographic differences in *Borrelia* infections with a higher prevalence in areas in Middle Europe.<sup>8</sup>

To further support this theory, Lyme disease affects mostly the skin: about 80% of all Lyme borreliosis cases present skin manifestations.<sup>15</sup> We recognise that classical dermatological events include erythema chronicum migrans (ECM), lymphadenosis benigna cutis (LABC) or borrellial lymphocytoma (BL) and acrodermatitis cronica et atrophicans (ACA), but there is growing evidence that some cases of other cutaneous manifestations could be related to borrellial late infection, mostly *Borrelia afzelii*.<sup>13</sup> The diagnosis of Lyme borreliosis is based on clinical data. Common laboratory tests are not usually revealing for the diagnosis of Lyme borreliosis and serologic tests support the diagnosis, but are not always essential in this regard.<sup>17</sup>

In this case of LSA mimicking scleroderma *en coup de sabre*, an association with Lyme borreliosis is proposed.

To our knowledge, this is the first case of LSA, mimicking scleroderma *en coup de sabre*, which was associated with Lyme borreliosis. Our findings indicate an association between this particular form of LSA and *Borrelia*, suggesting that *Borrelia burgdorferi* itself could represent a causative agent of this atypical form of LSA, however it cannot be excluded that *Borrelia* could be only one of the predisposing agent triggering it.

## Conclusions

In conclusion we propose that tissue PCR for DNA of *Borrelia* should be performed in patients with LSA in endemic area, because it could represent a rare manifestation of Borreliosis, and in those cases LSA should be treated with proper antibiotic therapy in order to eradicate the microorganism.

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