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Case Presentation

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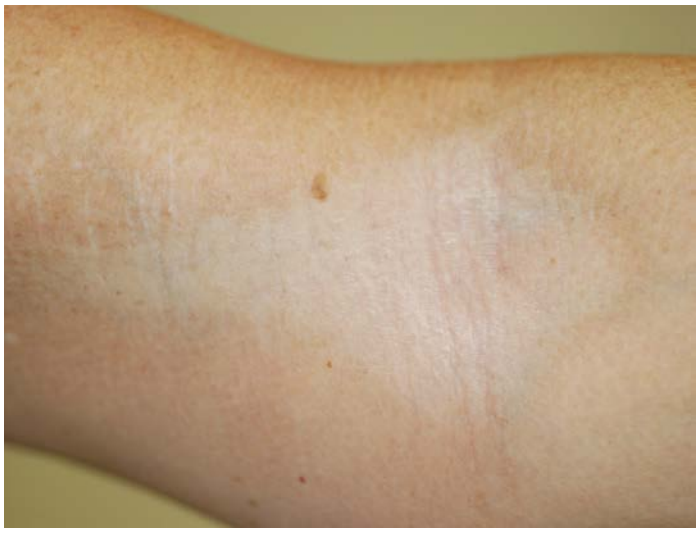
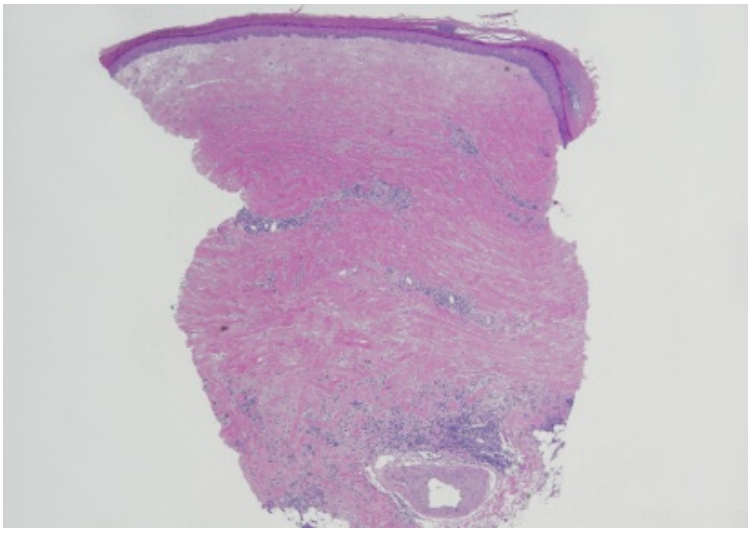
Abstract

Morphea and lichen sclerosus et atrophicus (LSA) have similar clinical presentations. Reports of patients with overlapping clinical and histopathologic features of both conditions have led some to speculate that they may represent different presentations along the same disease spectrum. It has been postulated that there is a common etiologic agent, which may involve autoimmunity, response to trauma, or infection. The link between *Borrelia* infection and both morphea and LSA has been widely studied but remains controversial. We present a case of a patient with lesions characterized by overlapping features of morphea and LSA with rapid decrease in joint mobility.





Case synopsis

A 69-year-old woman presented to the New York University Dermatologic Associates in June 2012, for evaluation of an eruption on her arms, abdomen, back, and inguinal folds that had been present for approximately three months. The lesions were associated with both pain and pruritus that interfered with her ability to sleep through the night. Treatment with antihistamines, fluocinonide, and a one-week course of prednisone failed to provide substantial relief of symptoms or clearance of the lesions. Over the next two months, the patient began to experience the progressive decreased range of motion in her arms.

Past medical history included thyroid cancer, hypertension, breast cancer, and Lyme borreliosis treated two years prior to presentation.

Physical Examination: There were large, diffuse, white, atrophic plaques on the flexural surfaces of the arms, the inguinal folds, the posterior aspect of the right shoulder and scapula, and the popliteal fossae. The plaques felt firm upon palpation and diminished the patient's range of motion.

Laboratory Data: The white -cell count was 13.4 x 10^{9}/L with a neutrophil count of 12,301 x 10^{9}/L. The total cholesterol and triglycerides were 226 mg/dL and 229 mg/dL, respectively, and the uric acid level was 7.9 mg/dL. Hepatitis panel was negative.

Histopathology: A punch biopsy specimen was obtained from the right groin. There is edema and sclerosis of the papillary dermis with telangiectases, mild epidermal atrophy, and hyperorthokeratosis. There is a superficial and deep, perivascular,

inflammatory infiltrate that is comprised predominantly of lymphocytes and scattered plasma cells with sclerosis and fibrosis throughout the deeper portions of the dermis.

Diagnosis: Lichen sclerosis et atrophicus and morphea

Discussion: Lichen sclerosus et atrophicus (LSA) most commonly presents as atrophic plaques in the genital region but can occur in extra-genital locations. The etiology remains unknown but has been hypothesized to be related to genetic susceptibility, autoimmunity, infectious agents such as spirochetes, and the Koebner phenomenon [1]. Morphea similarly presents with atrophic plaques and has also been postulated by some to be related to spirochete infection and autoimmune disease. In patients with generalized morphea, the plaques become large and confluent and involve more than two areas on the body. Although traditionally thought of as two separate entities, reports of patients with overlapping clinical and histopathologic features of both conditions have led some to speculate that they may represent different presentations along the same disease spectrum. They are sometimes both categorized under the broader heading of localized sclerosis owing to their similar clinical and histopathologic presentations; some have referred to LSA as subepidermal morphea [2].

Differentiating between morphea and LSA presents a clinical challenge because both conditions may present with white, sclerotic, indurated plaques. Patients with LSA, in contrast to those with morphea, more commonly experience intense pruritus [3]. Histopathologically, morphea is characterized by sclerosis of the reticular dermis, swollen collagen fibers, a perivascular infiltrate, and a loss of adnexal structures, whereas LSA shows follicular plugs and a lichenoid infiltrate in the papillary dermis. Some have proposed that the presence or absence of elastic fibers in the upper dermis may allow one to distinguish between the two conditions. Elastic fibers are normal in morphea but absent in LSA [3]. A retrospective study of 18 patients with LSA and 21 patients with morphea suggested that dermoscopy may yield some helpful diagnostic clues [4]. Dermoscopic features of LSA included comedo-like openings that corresponded to follicular plugs and white patches that were indicative of epidermal atrophy. The key feature of morphea was the presence of fibrotic bands that were thought to represent a sclerotic dermis. A single case report of a patient with overlapping LSA and morphea also found white structureless areas to be indicative of LSA [5].

A retrospective study of 472 patients with morphea showed that 5.7% had coexisting LSA on histopathologic examination [6]. The most common site of extragenital LSA in patients with morphea was the shoulders and only patients with plaque morphea or generalized morphea (as opposed to linear, guttate, deep morphea, or other subtypes) had coexisting LSA. A study of 76 patients with morphea showed that 38% also had genital LSA. The authors concluded that an examination of the genital region is warranted in morphea patients, owing to the possibility of evolution to SCC [7]. They further speculated that LSA is the genital manifestation of morphea.

Furthermore, morphea and LSA have both been related to *Borrelia* infection, with case reports of complete remission of LSA after treatment with antibiotics [8]. Our patient has a diagnosis of Lyme borreliosis, although the link between *Borrelia burgdorferi* infection and morphea or LSA remains controversial and is dependent upon geography. A study that utilized the polymerase chain reaction (PCR) to detect *Borrelia* DNA in archival, paraffin-embedded, skin specimens found that 2% of 49 morphea biopsy specimens and 6.6% of LSA biopsy specimens were positive for *Borrelia* [9]. They also compiled all of the 19 PCR-based studies performed prior to the date of publication in 2009, which showed that 11 studies failed to detect *Borrelia* DNA whereas the other eight had rates of positivity that ranged from 3 to100%. *Borrelia* has been detected in LSA in 50% of studies; in those showing detection, *Borrelia* DNA positivity rates ranged from 4 to100%. The wide range of positivity is likely a reflection of differences in the prevalence of *Borrelia* by geographic region. It is important to note that of the 99 skin biopsy specimens of patients from the United States, who were included in the literature review, there was only one case of *Borrelia* positivity, which occurred in an immunocompromised patient [9]. The rates of positivity, however, were as high as 18.2% in studies performed in Europe and Asia. The same general trend held true for *Borellia* positivity in specimens of LSA.

Trauma and previous inflammation also have been proposed as an etiology for both morphea and LSA, particularly for patients that have overlapping features of both conditions. One case report of a patient with overlapping LSA and morphea was thought to represent an isotopic response to a prior zoster infection in the same area [10]. An isotopic response is the presence of a new skin disease at the site of a different, unrelated, previously- healed disease. The patient developed thick, gray, sclerotic, and pruritic plaques that were localized to the C4 to C7 dermatome on the left side one month after a case of healed zoster [10]. LSA not uncommonly occurs after trauma, such as radiation or infection.

Treatment options for both LSA and morphea include topical glucocorticoids, antimalarials, colchicine, methotrexate, and phototherapy. Several studies have established the efficacy of ultraviolet A for the treatment of localized morphea and case reports suggest that narrow-band ultraviolet B may also halt disease progression [11]. Although highly effective for morphea, phototherapy is considered to be less effective for LSA. Patients with generalized morphea have much larger plaques than do those with localized disease and often require more aggressive treatment because of the risk of joint contractures with subsequent disability. For patients with rapidly progressive disease, many favor the combination of methotrexate with glucocorticoids. One

case report of a patient with what appeared clinically to be generalized morphea but with histopathologic features of both LSA and morphea had an excellent response to sulfasalazine [12]. After one month, the mobility of the affected joints was completely restored.

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