

CASE REPORTS

Patient With Generalized Guttate Morphea and Lichen Sclerosus et Atrophicus

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Abstract. Generalized guttate morphea is a very uncommon clinical entity, and few reports are available in the literature. We report the case of a 7-year-old boy who first attended our clinic in 1990 with guttate morphea on the trunk and upper limbs. These lesions were associated with plaque morphea on his right foot. Twelve years later, lesions with a different appearance to the previous ones were observed in the right pectoral region. Clinically and histopathologically, they resembled lichen sclerosus et atrophicus. Given that morphea and lichen sclerosus et atrophicus share certain clinical and pathologic characteristics, some authors believe that these entities may be related or even different presentations of the same disease. The most noteworthy aspect of our case is the type of morphea, as we were unable to find equivalent examples in the literature.

Key words: guttate morphea, plaque morphea, scleroderma, lichen sclerosus et atrophicus.

COEXISTENCIA DE MORFEA EN GOTAS GENERALIZADA Y LIQUEN ESCLEROATRÓFICO: A PROPÓSITO DE UN CASO

Resumen. La morfea en gotas generalizada es una entidad clínica muy poco frecuente, con escasas referencias en la literatura. Describimos el caso de un varón de 7 años que acudió por primera vez a nuestra consulta en 1990 presentando este tipo de morfea en el tronco y las extremidades superiores, asociado a la variante en placas en el pie derecho. Doce años después le aparecieron unas lesiones en la región pectoral derecha de aspecto diferente a las previas, y que resultaron ser características clínica e histopatológicamente de liquen escleroso y atrófico (LEA). La morfea y el LEA comparten algunas características clínico-patológicas, y por ello algunos autores creen que podría tratarse de entidades emparentadas, e incluso de dos presentaciones distintas de una misma enfermedad.

Creemos que lo más llamativo del caso que describimos es el tipo de morfea que presenta, de la que no hemos encontrado ejemplos equiparables en la literatura.

Palabras clave: morfea en gotas, morfea en placas, esclerodermia, liquen escleroatrófico.

Introduction

Generalized guttate morphea is a chronic inflammatory disorder that has been rarely reported in the literature. It is a superficial form of scleroderma without systemic

involvement; it presents with multiple, small, infiltrated plaques measuring 2 mm to 10 mm that are normally violaceous in color, although on occasions they may be whitish. Guttate morphea typically follows a self-limited course after an initial progressive phase. The lesions tend to be localized and normally occur in combination with larger plaque-type lesions. There have been very few reports published on generalized guttate morphea.

Lichen sclerosus et atrophicus (LSA) is another chronic inflammatory dermatosis of unknown origin; it typically affects the genital area, although, as seen in our patient, it may also occur on other parts of the body.

It presents with small, shiny plaques that are whitish in color and have an atrophic (cigarette paper) appearance. The lesions often coalesce to form larger plaques.

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Figure 1. Morphea plaque on heel.

Case Description

We present the case of a 7-year-old boy who first visited our clinic in 1990 with a lesion on the right heel, followed by the appearance of numerous small lesions scattered across the trunk and upper limbs a few weeks later. There was no personal or family history of interest and the boy was not on medication at the time. The patient's weight and height were normal for his age and he reported no systemic symptoms.

Physical examination revealed a hard brownish plaque measuring 3 cm in diameter on the heel of the right foot (Figure 1); the plaque was depressed in comparison with the normal surrounding skin. We also observed 2 similar lesions on the dorsum of the same foot and multiple flat lesions, measuring under 4 mm, on the abdomen, upper arms, and anterior aspect of the forearms (Figure 2), and on the chest and upper back (Figure 3). The lesions were darker than the surrounding healthy skin, slightly infiltrated on palpation, and in some cases had coalesced to form larger plaques. They were also slightly depressed and asymptomatic.

The following tests were requested: routine blood and urine tests, protein electrophoresis, immunoglobulin and complement concentrations, and serological tests for *Borrelia*, antinuclear antibodies (ANAs), anti-DNA antibodies, and anticentromere antibodies (including anti-Scl-70 antibodies).



Figure 2. Guttate morphea lesions on the right forearm and inner aspect of the upper arm. Note how these lesions coalesce on the forearm.

The only pathological findings were positive ANA titers (1:100). A biopsy of the heel lesion revealed normal epidermis, fibrosis of the reticular dermis that extended to the subcutaneous cellular tissue, and sweat gland atrophy. All of these changes were consistent with a diagnosis of morphea. A biopsy of 1 of the lesions on the trunk revealed mild fibrosis, a minimal perivascular inflammatory infiltrate, and a reduced number of appendages, suggesting superficial morphea. On the basis of the above clinical and pathological findings, we established a diagnosis of generalized guttate morphea.

In the 16 years the patient has been followed up in our clinic, he has received treatment with topical corticosteroids, keratolytic agents, intramuscular penicillin G benzathine, synthetic antimalarial agents (hydroxychloroquine), and heliotherapy. During this time, the lesions have remained

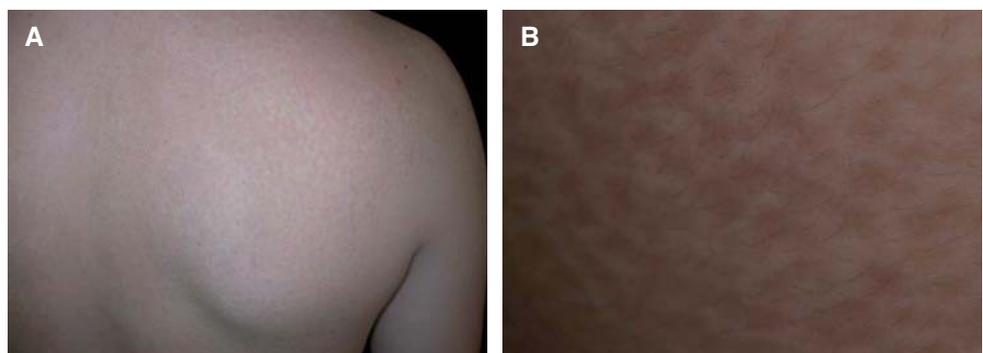


Figure 3. A, guttate morphea lesions on the back. B, more detailed view of back (the lesions in question are the small, slightly depressed brownish plaques).



Figure 4. Guttate morphea lesions on the chest. The 2 whitish plaques in the right pectoral region are suggestive of lichen sclerosus et atrophicus.

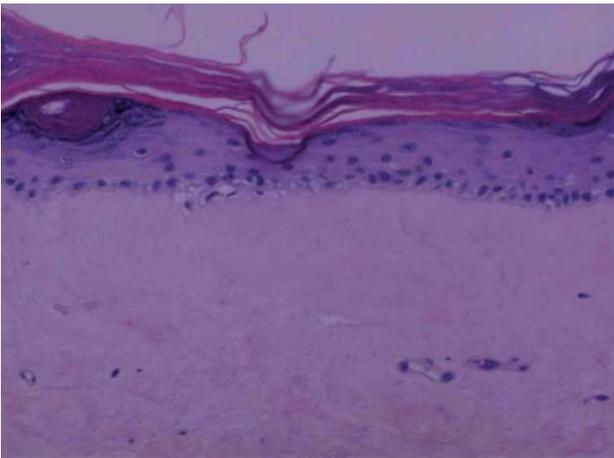


Figure 5. Biopsy of lesion in the right pectoral region. Note the hyperkeratosis, epidermal atrophy, hyalinization of the papillary dermis, and focal vacuolar degeneration of the basement layer (hematoxylin-eosin, original magnification $\times 400$).

stable and he has not developed dysphagia, arthralgia, or Raynaud phenomenon.

In October 2004, the patient presented with 2 asymptomatic lesions of recent onset in the right pectoral region (Figure 4). These lesions differed from the others: they were of a whitish-ivory color, noninfiltrated, had an atrophic (cigarette paper) appearance with small ecchymoses on their surface, and measured approximately 1 cm in

diameter. A biopsy of 1 of the lesions revealed a thinned, flattened epidermis with hyalinization of the superficial dermis and areas of hydropic degeneration of the basement membrane, accompanied by a minimal periadnexal inflammatory infiltrate (Figure 5). These findings, together with the clinical appearance of the lesions, suggested a diagnosis of LSA. The plaques have remained stable since their onset and no new lesions have appeared.

Discussion

Morphea, the most common form of scleroderma, is characterized by limited involvement of the skin, subcutaneous tissue, and occasionally, underlying muscle. Of the many clinical variants that have been described, plaque-type morphea is the most common.¹ It mostly affects women aged between 20 and 40 years but 15% of patients develop it before the age of 10 years.² Lesions develop insidiously as plaques of varying sizes that are indurated on palpation and occasionally surrounded by an erythematous-violaceous halo known as a lilac ring. This ring is a manifestation of inflammatory activity and, therefore, a marker of lesion progression.³ Disease progression varies according to the extent of the sclerosis; there are superficial forms with a benign course and minimal infiltration, and other, more severe forms affecting deeper structures and with the potential to cause serious disability.⁴ In most cases, however, with time, the lesions tend to stabilize in terms of size and number and the skin also becomes less sclerotic, although color changes persist to some extent in affected areas. Very few cases of guttate morphea have been reported in the literature and most of these have been associated with plaque morphea. Guttate morphea lesions, however, tend to be less infiltrated on palpation and to appear at an earlier age.

LSA is a chronic inflammatory disease that is most common in women and that involves the genitalia in 85% of cases. The lesions present as flat, shiny, bluish-white papules that, with time, coalesce into larger plaques with an atrophic, wrinkled appearance. While extragenital LSA is less common (15% of cases), it has been more frequently reported in association with morphea.^{5,6} In such cases, the lesions tend to affect the upper part of the trunk, the neck, the arms, and areas that have been subject to continuous pressure or trauma.

While LSA and morphea are of unknown etiology, they have both been associated with numerous autoimmune disorders such as systemic lupus erythematosus,⁷ Hashimoto thyroiditis,⁸ and diabetes mellitus.⁶ They have also been related to *Borrelia burgdorferi* infection, although results have varied between populations.^{9,10} In addition, high titers of ANAs and anti-DNA antibodies are often found in some generalized forms of morphea.¹¹

There have also been relatively frequent reports of morphea and LSA lesions in graft-vs-host-disease,¹² supporting the theory that an autoimmune mechanism is involved in both processes. The literature contains many reports of an association between LSA and morphea, although this relationship is controversial.¹³ Patterson and Ackerman,¹⁴ for example, do not believe in such an association. In 1984, they stated that to distinguish between the 2 diseases, it was necessary to analyze the reticular dermis and adipose tissue, regardless of changes observed in the epidermis or papillary dermis. In their opinion, deeper structures are affected by morphea but not by LSA. Furthermore, in 1991, Shono et al,¹⁵ reported that staining patterns for the 2 disorders were different, supporting the idea of 2 different diseases.

Other authors, in contrast, believe that there is evidence to support an association between morphea and LSA. In 1980, for example, Uitto et al¹ described the co-existence of the 2 types of lesions—both clinically and histologically—in a series of 10 patients. Also in favor of this theory are Connelly and Winkelmann,⁵ who stated that LSA was a morphea of the papillary dermis; thus, depending on the level at which sclerosis occurred, there could be 3 types of lesion: LSA, typical morphea, and subcutaneous morphea. The coexistence of the 2 types of lesions in our patient also suggests that the 2 processes are closely related.

In conclusion, we believe that the association described between LSA and morphea in our patient is worthy of note but what is more remarkable, in our opinion, is the unusual form of morphea identified in our patient, particularly because we were unable to find equivalent examples in the literature.

Conflicts of Interest

The authors declare no conflicts of interest.

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