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Coexisting Morphea and Extragenital Lichen Sclerosus ET Atrophicus in a Female Patient

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Abstract
Morphea and lichen sclerosus et atrophicus (LSA) are two entities that are characterized clinically by indurated and sclerotic plaques with overlying dyschromic skin and pathologically by an inflammatory dermal infiltrate and fibrosis. The exact relationship between morphea and LSA is debateful. Some authors consider LSA a superficial variant of morphea whereas others consider them as two unrelated entities. Here we present a case of co-existing plaque morphea and linear morphea with concomitant lesions of extra genital LSA occurring at the same site in an adult female.
Keywords- *Morphea, lichen sclerosus et atrophicus, sclerosis, plaque.*

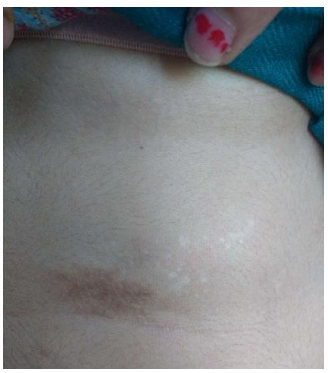
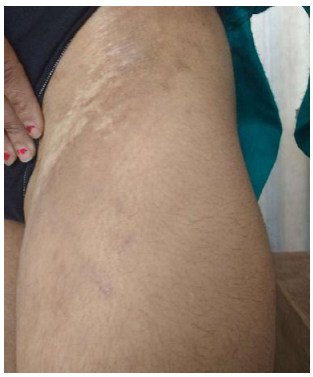
INTRODUCTION

Morphea or localized scleroderma is an inflammatory disease of the skin and sub cutis that leads to sclerosis of the underlying tissues. The incidence varies between 0.34 and 2.7 cases per 100,000 populations per year. Plaque morphea is the most common variant of localized scleroderma. Linear morphea or linear scleroderma is the most common variant of morphea in the childhood which presents with a single unilateral lesion distributed mostly on the limbs, face, or scalp. Clinically the lesions of morphea appear as poorly defined plaques often depressed with alterations of pigmentation[1]. Lichen sclerosus et atrophicus (LSA) was first described by Hallopeau in 1887. It is a benign chronic inflammatory dermatosis affecting both the epidermis and the dermis. Typical clinical findings are ivory white opalescent clustered papules which progressively result in parchment like skin. LSA occurs mainly in the anogenital area (83% to 98%) and sometimes at extra genital sites (15% to 20%). Here we present a case of co-existing plaque morphea and linear morphea with concomitant lesions of extra genital LSA occurring at the same site in an adult female[2].

Case Report

A 27-year-old female presented with a 1-year-old history of an asymptomatic hyper pigmented non progressive plaque over the abdomen of size of a

palm. 4 months ago, she noticed a slowly expanding band of dyschromia associated with tightness over the affected skin extending from the inguinal fold upto the calf. On examination, there was a hyper pigmented indurated plaque over the abdomen of size 5×4 cm. Adjacent to the plaque were multiple, well defined discrete to confluent ivory white macules of size 1-2mm with surface atrophy. Over the right lower limb, there was a band of indurated plaque extending from right inguinal fold along the medial part of the thigh upto the calf posteriorly with a width of 5-7 cm. The surface showed atrophy with areas of hyper and hypo pigmentation and loss of appendages. At the upper end of the lesion and the posterior thigh overlying the plaque, there were discrete grouped white papules of size 1-2 mm with surface atrophy. Underlying joint movements were normal. Systemic examination was non-contributory. The biopsy from the plaque showed homogenized thick bundles of collagen extending to deep dermis and subcutis with pulling up of eccrine coils and mild perivascular lymphoplasmacytic infiltrate. The biopsy from the popular lesions showed flattening of the epidermis with loss of normal rete pattern with thickening dermis, hylanised pale collagen and paucity of adnexal structures. On the basis of clinical and histopathological examination, we inferred that patient had coexistence of morphea and extra genital lichen sclerosus et atrophicus. The patient was put on topical calcipotriol and betamethasone ointment.

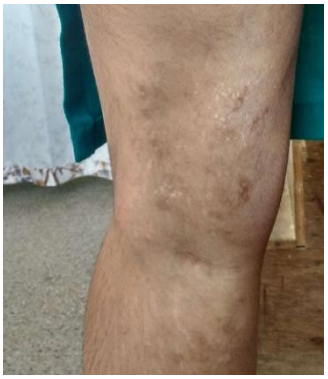
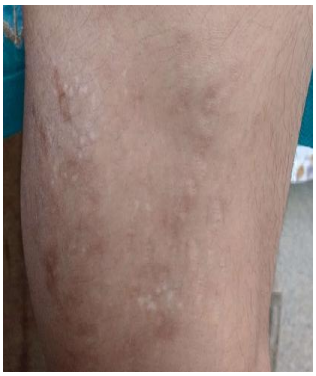



Figure 1-4 showing lesions of LSA overlying the lesions of morphea

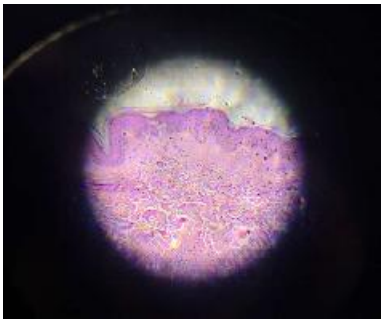


Figure 5 showing histopathological image (10x).

Discussion

Morphea and lichen sclerosus et atrophicus are two entities that are characterized clinically by indurated and sclerotic plaques with overlying dyschromic skin and pathologically by an inflammatory dermal infiltrate and fibrosis. Etiology is largely unknown and both genetic factors, such as predisposing HLA alleles, and environmental factors, such as infection with *Borrelia burgdorferi*, have been implicated. Autoimmune diseases are more frequent in patients with morphea or LSA than in unaffected persons. The exact relationship between morphea and LSA is debateful. Some authors consider LSA a superficial variant of morphea whereas others consider them as two unrelated entities. In some cases, referred to as "white spot disease," the differentiation between morphea and LSA can be impossible[3]. Uitto et al[4] were the first to describe a series of 10 patients in which the two entities coexisted. Histopathological study of these cases indicated features of both morphea and LSA with equal intensity in seven of them. Subsequently, sporadic cases of these two coexisting diseases have been published. Chen et al[5] reported a case of co-existing morphea and LSA

occurring in a zosteri form pattern exhibiting positive treatment response to plaquenil and topical steroid. Blaya et al[6] reported a patient with guttate morphea who developed lichen sclerosus et atrophicus lesions after twelve years. In the study by Uitto et al[4], repeated biopsies were taken from patients at the same location at varying time points. In one case, a transition from LSA into morphea was found over a period of 2 years, with complete disappearance of LSA eventually[4]. Wallace[7] studied 380 LSA patients and concluded that histologically-confirmed morphea was present in 13 of these patients. Schaffer et al[8] described six patients with both LSA and morphea as manifestations of sclerodermoid chronic graft-versus-host disease. In one of the six patients, the morphea form plaques underwent a transition to whitish, superficial LSA like lesions before resolving. A revised classification of morphea to include LSA as a subtype owing to the histologic similarities between the two conditions has also been proposed[5].

However, many investigators have argued against the coexistence of morphea and LSA and insist that there is sufficient clinical and histologic difference between them. A discriminating method using elastic tissue staining has been devised and it has been inferred that elastic fibers in the upper dermis are lost in LSA, but not in morphea. Kowalewski et al[9] used laser scanning confocal microscopy and proposed that the alteration of the basement membrane zone in morphea was different from that in LSA. The former preserved continuity of structures in basement membrane zone, which was lost in the latter. Our case presented simultaneous occurrence of both types of clinical manifestation which proved to be morphea and LSA by histopathology. Partial improvement was noted with the treatment after two months.

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