An unusual presentation of localized bullous morphea

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Abstract

Bullous morphea is a rare variant of localized scleroderma characterized by occasional intermittent blisters. Lichen sclerosus is a chronic inflammatory disease. The coexistence of morphea and lichen sclerosus has been reported in different sites in the same patient and more rarely in the same lesion. We report the case of a 54-year-old woman with an atypical presentation of bullous morphea and some histological features of lichen sclerosus. She presented with a 5-year history of an ulcerated plaque, with a sclerotic and atrophic center and indurated budding margins, localized on the lumbar back. Initially the diagnosis of a squamous cell carcinoma was suggested. A skin biopsy confirmed the diagnosis of bullous morphea and showed some histological features of lichen sclerosus. Topical betamethasone and silicone gel ointment were prescribed leading to complete healing of the ulceration within five months. Our case is unusual because of the atypical clinical presentation, the histological aspect combining signs of bullous morphea and lichen sclerosus, and the favorable results with the use of local corticotherapy and silicone gel.

Keywords: morphea, bullous, lichen sclerosus, silicone therapy

Introduction

Morphea, also called localized scleroderma, is a sclerosing condition of the skin and subcutaneous tissue. Morphea has been classified into 5 subtypes: plaque or circumscribed, linear, generalized, bullous, and deep (including subcutaneous morphea), [1]. Bullous morphea is rare. To the best of our knowledge, only 67 cases have been reported in the literature [2, 3]. Coexisting cases of morphea and lichen sclerosus have been reported rarely in the same plaque. We report herein a case of a long-standing ulcer on the back from which skin biopsy revealed histological aspects of bullous morphea with features of lichen sclerosus. The patient was treated successfully by local corticosteroid therapy and silicone gel.

Case Synopsis

A 54-year-old woman with a medical history of hypertension, diabetes mellitus type II, and hypothyroidism, consulted our dermatology department for a 5-year history of ulcerated plaque on the back. The patient had a surgical history of

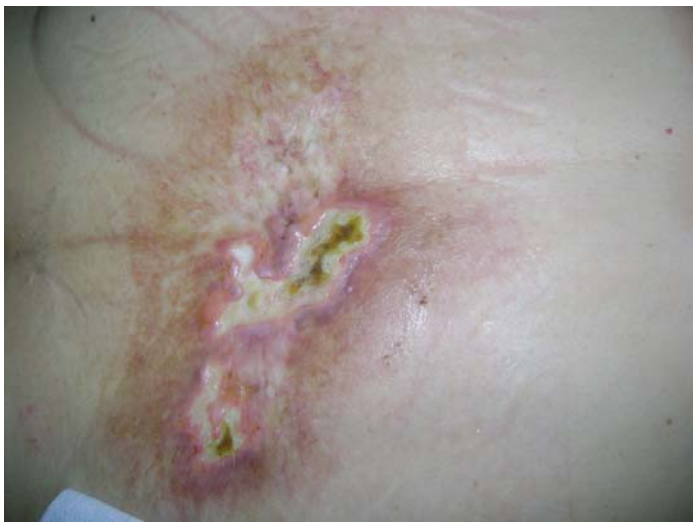

Figure 1. *Sclerotic and ulcerated plaque on the back.*

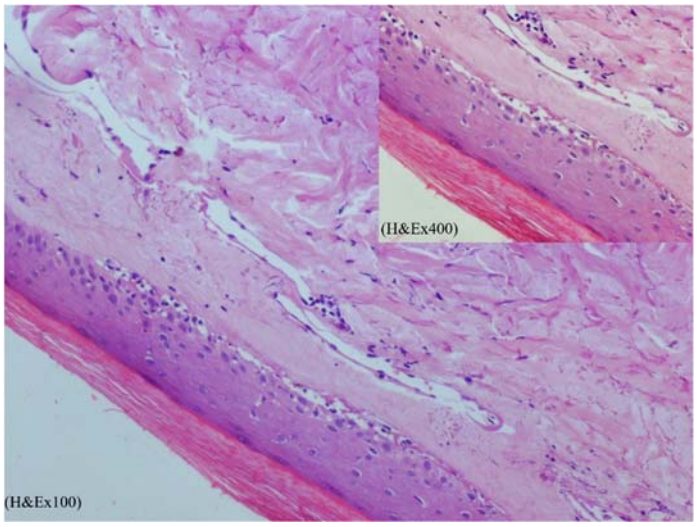

Figure 2. *Lymphoplasmocytic infiltrate at the dermal-epidermal junction associated with thickened collagen bundles in the reticular dermis. H&E, 40×.*

cervical and lumbar laminectomy 15 years prior. On physical examination, there was a 15×10cm sclerotic erythematous, telangiectatic, and ulcerated plaque with atrophic areas (**Figure 1**). No lesions were present in the genitalia and oral mucosa. She had no other symptoms. Initially, the diagnosis of cutaneous squamous cell carcinoma was considered.

A skin biopsy revealed atrophic epidermis. The dermis showed typical features of morphea with collagen hyalinization, thickened collagen bundles in the reticular dermis, and associated lymphoplasmacytic infiltrate at the dermo-epidermal junction (**Figure 2**). Lymphangiectasia

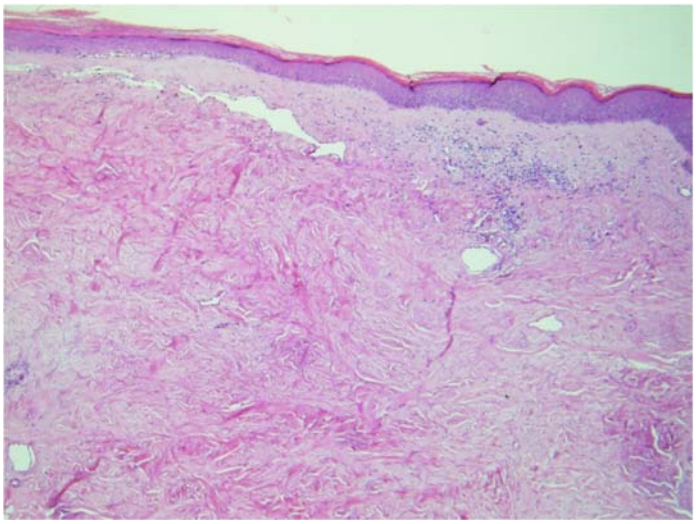

Figure 3. *Atrophic epidermis, dilated lymphatic channel in the dermis and focal* basal *cell* vacuolization. *H&E, 100×.*

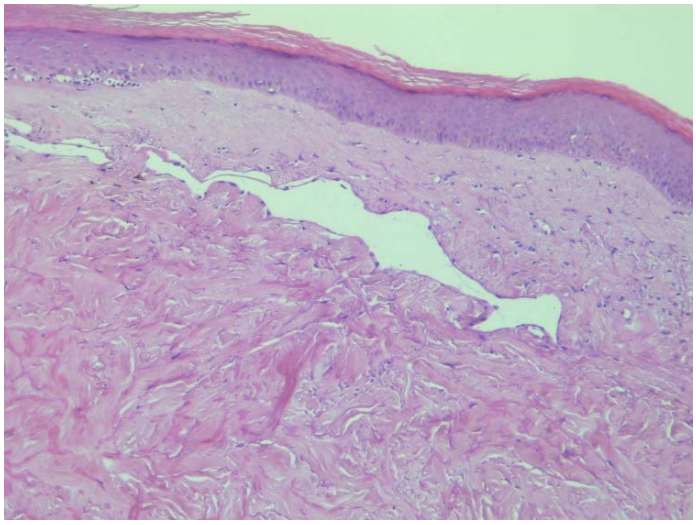

Figure 4. *Edema of the papillary dermis and a loss of elastic fibres in the superficial dermis indicated by orcein stain. H&E, 40×.*

was present (**Figure 3**). There was focal *basal* cell *vacuolization* (**Figure 3**), edema of the papillary dermis, and a loss of elastic fibers in the superficial dermis indicated by orcein stain (**Figure 4**). The diagnosis of bullous morphea with some histological features of lichen sclerosus was determined. Complementary tests were normal. Treatment with topical corticosteroid and silicone gel led to the resolution of the ulceration and inflammatory component within five months (**Figure 5**).

Case Discussion

Bullous morphea is a rare variant of localized scleroderma. Its frequency ranges from 1%, 4 to 7%,

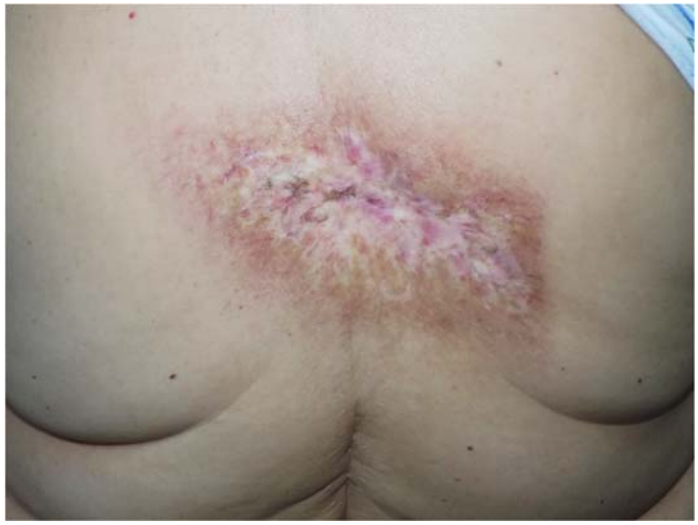

Figure 5. *Healing ulceration on the back.*

and 5% of all cases of scleroderma [3, 4]. Bullous morphea is frequently located on the lower extremities [5]. Lichen sclerosus (LS) is an inflammatory disorder with a mainly genital location. Extragenital bullous LS is rare. Bullous lesions are usually transient and can be hemorrhagic and heal before the appearance of typical plaques, sometimes with milia formation [6].

In our case, histopathologic examination showed an inflammatory dermal infiltrate, thickened collagen bundles in the reticular dermis, and lymphectasia, all of which are consistent with the diagnosis of bullous morphea. Also, some histological features of LS were present, such as focally *basal* cell *vacuolization* and loss of elastic fibers in the superficial dermis indicated by orcein stain [4]. Other characteristic aspects of LS like hyperkeratosis and follicular plugging were absent in our case.

Morphea and LS are both considered autoimmune phenomena [7]. In their typical form, the two diseases are often distinguishable clinically and histologically. Some authors suggest that they are a part of the same spectrum and consider LS a subepidermal morphea. Cases of co-existence of morphea and LS have been reported in the same patient. There have been reports of morphea/LS overlap [8, 9]. The edema seen in LS is attributed to lymphatic obstruction related to deep sclerosis in the morphea plaque [8]. Lymphatic dilatation is incriminated in the development of the bullae also in bullous morphea. However, it is not a constant finding [5, 10].

Traumatic origin has been reported as a trigger factor in the two diseases. Minor trauma may engender blister formation in extragenital LS. Cases of bullous morphea, bullous LS, and overlap morphea/LS have been reported after radiation therapy [7, 11]. Lichen sclerosus has been reported after mastectomy, and exposure to welding sparks [12]. In our case, lumbar laminectomy could be considered as a potential cause of bullous morphea in this location.

The treatment of bullous morphea is difficult, especially in its ulcerated form. Systemic and intralesional corticosteroids and antimalarial agents have been reported in the treatment of bullous morphea [5]. A complete healing course of the ulcer was noted in our case after applying topical corticosteroid and silicone gel. Local corticosteroid therapy is usually prescribed for morphea and lichen sclerosus. Silicone gel is not a classic treatment of the two diseases. In fact, a case of silicone-induced morphea after breast implant was reported [8, 13]. However, this is an internal form of silicone. The mechanism of action of silicone-induced morphea is not clear. Silicon injection in soft tissue leads to a perivascular lymphocytic infiltration with small local deposits of immunoglobulins around the walls of small vessels and subsequently a fibroblastic reaction in the skin.

However, in our case, the topical silicone gel had a treatment effect that could be explained by its mechanisms in the treatment of hypertrophic scar. The authors suggest various hypotheses such as increasing oxygen tension or temperature, direct action of silicone oil, polarization of scar tissue caused by negative static charge, and modulation of growth factors [9, 14, 15].

Conclusion

Our case was interesting in its clinical presentation of a long-standing ulcerated plaque, its histological aspect combining features of bullous morphea and LS, and the favorable results with the use of local corticosteroid therapy and silicone gel.

Potential conflicts of interest

The authors declare no conflicts of interests.

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