An unusual case of combined bullous Lichen sclerosus et atrophicus and morphea disease

Maryam Sadat Sadati[1], roya radanfar[2], razieh ahmadi[2], and Fatemeh Sari Aslani[3]

[1]molecular dermatology research center ,Department of dermatology ,school ofmedicine ,shiraz university of medical sciences, shiraz
[2]Shiraz University of Medical Sciences
[3]3- Department of Pathology, School of Medicine, Shiraz University of Medical Sciences, Shiraz, Iran

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Abstract

LSA and morphea are chronic diseases, characterized by the sclerosis of the connective tissues. Bullous type of both diseases and coexistence of LSA and morphea are very rare manifestations. Herein. we report a case consistent with bullous combined LSA and morphea that was treated with methotrexate and pulse of methylprednisolon

An unusual case of combined bullous Lichen sclerosus et atrophicus and morphea disease

Maryam Sadat Sadati[a,b,*], Roya Radanfar[ab] Razieh Ahmadi [ab], .Fatemeh Sari Aslani [c]

[a]Molecular Dermatology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

[b]Department of Dermatology, Shiraz University of Medical Sciences, Shiraz, Iran

[c]Assistant professor of Pathology, Pathology Department, Shiraz University of Medical Sciences

*Corresponding author: Msadati63@yahoo.com

Abstract

Aim : The present report describes a case of combined bullous Lichen sclerosus et atrophicus (LSA) and morphea which is a rare finding.

Summary: LSA and morphea are chronic skin diseases, characterized by the sclerosis of the connective tissues. Bullous type of both diseases and coexistence of LSA and morphea are very rare manifestations.

Herein. we report a case consistent with bullous combined LSA and morphea that was treated with methotrexate and pulse of methylprednisolone

Key learning points:

- Bullous types of LSA and morphea is very rare .
- Coexistence of LSA and morphea is a very rare finding .
- LSA and morphea are different chronic inflammatory skin diseases but they are on the same spectrum .
- Bullous LSA can be mimicked by bullous morphea .**Keywords:** lichen sclerosus et atrophicus, morphea,bullous LSA,

Consent statement : Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Introduction

Lichen sclerosus et atrophicus (LSA) and morphea are chronic inflammatory skin diseases with unknown etiology, characterized by sclerosis of the connective tissues. Bullous type of both diseases is a rare finding (1-4). LSA usually occurs alone, but the coexistence of lichen sclerosus et atrophicus and morphea is a rare finding. Several authors believe that these two diseases have different manifestations. but they are on the same spectrum (1, 2, 5).

Herein, we report a case consistent with bullous combined LSA and morphea that was treated with methotrexate and methylprednisolone pulse.

Case report

A 67-year-old woman referred to dermatology ward. She had positive history of hypertension, hyperlipidemia and eczema due to pruritus and blistering lesions on the anterior trunk and inguinal area since four months prior to being admitted. Clinical examination revealed generalized skin dryness, erythematous, slightly indurated plaques containing bullae on the anterior trunk and inguinal area, dull erythematous and sclerotic plaques on both legs (Figure 1,2). Two punch biopsies were taken from the trunk and leg. Microscopic examination of the trunk lesion showed hyperkeratosis, thinning of epidermis, edema of papillary dermis with focal subepidermal bulla containing RBCs, focal mild lymphocytic infiltrate in the mid and deep dermis and homogenization of collagen fibers of the reticular dermis. Microscopic examination of leg lesion showed hyperkeratosis, mild thinning of epidermis, homogenization of collagen fibers, mild perivascular and focal interstitial lymphocytic infiltrate in the superficial and deep dermis. According to the mentioned histopathologic report, simultaneous features of both bullous LSA and morphea was seen in the same lesion of trunk and features of morphea alone was seen on the leg lesions (Figure 3,4). Methotrexate 7.5 mg orally was prescribed. Due to incomplete response pulse of methylprednisolone 250 mg intravenously in 3 consecutive days was added. Both types of lesions were treated successfully.

Discussion

Our patient was presented with sclerotic plaques on her trunk and lower extremities with coexistence of bulla on the sclerotic plaques on the trunk with diagnosis of concurrent bullous LSA and morphea in the same lesion.

Simultaneous bullous LSA and morphea is a very rare disease. Morphea and LSA can coexist in a same patient and in spite of different clinical and histomorphological presentations, simultaneous occurrence of both conditions in a single lesion indicates overlapping similar possible etiopathogenesis (1).

However, it is not very clear as to why bullous morphea and LSA coexist. This close relationship raises the question of whether morphea and LSA are different manifestations of the same disease (2).

Abhijit Das and his colleagues reported a 4-year-old girl presented with a nonpruritic hypopigmented sclerotic patch over her left shoulder that histopathological examination showed features consistent with both LSA and morphea in the same lesion (1).

Sirin Yasar reported a 70-year-old man presented with annular, ivory colored, atrophic plaques, surrounded by erythema on his trunk with occasional bulla formation on the plaques for 6 months. Histopathologic examination was consistent with bullous morphea and LSA (2).Bullous morphea usually characterized with bullae on or around atrophic morphea plaques .Cllinicaly, bullous morphea may resemble Bullous Lichen sclerosus and the clinical setting is insufficient to make the diagnosis. However, they are differentiated by histological examination and only bullous LSA shows hyperkeratosis, follicular plugging, or epidermal atrophy with vacuolar change of basal cells, which were seen in our case.

Another study by Ardian cuellar-barboza described a 56-year-old woman with progressive bullous sclerotic lesions diagnosed as bullous morphea (6).

Can Baykal reported a 74-year-old woman with a 6-month history of multiple asymptomatic, shiny, indurated plaques located on her abdomen and back with diagnosis of morphea-LSA overlap (5).

Conclusion

Although coexistence of LSA and morphea is very rare, in every patient with overlapping clinical findings of morphea and LSA, the possibility of both diseases should be kept in mind.

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Conflict of interest: None

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Detailed author"s contribution :

Conception and design:Maryam Sadat Sadati

Drafting or revising the article:Roya Radanfar,Razieh Ahmadi

Provided grammatical revision :Roya Radanfar

Investigaton :Fatemeh Sari Aslani

Provided revisions to scientific content of manuscript: Maryam Sadat Sadati,

Data availability statement: the dataset generated during the current study are available from the corresponding author on reasonable request.

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Figure legends:

Figure 1: erythematous plaques

Figure 2: erythematous, indurated plaques with occasional ruptured bulla

Figure 3: hyperkeratosis, thinning of epidermis, edema of papillary dermis with focal subepidermal bulla and homogenization of collagen fibers of the reticular dermis.

Figure 4: hyperkeratosis, mild thinning of epidermis, homogenization of collagen fibers