Case Report

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Extensive isolated extragenital hemorrhagic bullous lichen sclerosus in a 75-year-old Yemeni woman: A case report

Volume 8 Issue 1 - 2024

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Received: March 02, 2024 | **Published:** March 15, 2024

Keywords: hemorrhagic, extragenital, lichen sclerosus et atrophicus, bullous

Abbreviations

LS, lichen sclerosus; EGLS, extragenital lichen sclerosus; EGBLS, extragenital bullous lichen sclerosus; BLSA, bullous lichen sclerosus et atrophicus

Introduction

Lichen sclerosus (LS) is a rare inflammatory mucocutaneous dermatosis described first by Hallopeau (1887) and later histopathologically by Darier (1892).[1] It primarily affects the anogenital skin (85%), and rarely, the extragenital skin (15%).[2] LS had different names until 1976, when Friedrich proposed the removal of "et atrophicus" from "lichen sclerosus et atrophicus." This is because LS is not histologically atrophic in all cases. Thereafter, the term "lichen sclerosus" was adopted.[2] The bullous clinical variant of LS, caused by the hydropic degeneration of basal keratinocytes, is rare. The hemorrhagic bullous variant is even rarer.[1]

While LS affects both sexes and all age groups, its incidence is higher in women than in men (10:1), especially among those aged 50–60 years.[2] The anogenital area is predominantly involved, with an extragenital-to-anogenital lesion ratio of 1:5.[3] Exclusive extragenital LS (EGLS) without genital lesions is extremely rare, whereas EGLS with concomitant genital involvement is common. Both can be classified as localized or disseminated depending on the surface area. Only 45 cases of EGLS with bullous lesions (EGBLS) have been reported, comprising 32 and 13 cases of exclusive EGBLS and EGBLS with concomitant genital lesions (Table 1), respectively. The 32 EGBLS-only cases comprise 10 and 22 cases of hemorrhagic and non-hemorrhagic bullae, respectively. Only two exclusive EGBLS cases with disseminated hemorrhagic lesions have been reported; this report describes the third case to date.[1] Clinically, extragenital lesions present as whitish papules and plaques with wrinkled surfaces and superficial horny plugs.[4] They are seldom pruritic but are usually asymptomatic. Herein, we report a case of extensively isolated extragenital bullous and hemorrhagic LS, an atypical and extremely rare variant of the disease.

Table 1 Summary of the reported cases of bullous EGLSA

Serous/ hemorrhagic	Localised/ disseminated	Exclusive EG/with G	Site	Number	Sex	Age	Title	1st Author	Date of publication	
H	D	EG	neck, arms, back, flank, chest	1	M	64	Generalized bullous and hemorrhagic lichen sclerosus et atrophicus. Marked improvement with ACTH	Di Silverio A	1975	1
S	D	EG	abdomen, breast, axilla, chest, thigh	1	F	55	Extensive lichen sclerosus et atrophicus with bullae and ulceration improvement with hydroxychloroquine	Wakelin SH	1994	2
S	D	EG	scalp, trunk, upper limb	1	M	84	Extensive bullous lichen sclerosus with scarring alopecia	Madan V	2009	3
S	D	EG	anterior neck, trunk, arms, forearms, thigh, flank, abdomen	1	F	61	Disseminated extragenital bullous lichen sclerosus	Trinh TVT	2014	4

Table 1 Continued...

Serous/ hemorrhagic	Localised/ disseminated	Exclusive EG/with G	Site	Number	Sex	Age	Title	1st Author	Date of publication	
S	D	EG	Back, arms, forearms, and medial thighs	I	F	61	A New Case of a Disseminated Extragenital Bullous Lichen Sclerosus	Amarouch H	2016	5
H	D	EG	lower Back, Buttock, abdomen	I	F	63	Extensive bullous lichen sclerosus et atrophicus	Vukicevic J	2016	6
S	D	EG	Back, lower limb	I	M	35	Bullous lichen sclerosus-generalized morphea overlap syndrome improved by tofacitinib	Liu L	2022	7

Abbreviations: D: disseminated, EG: exclusive extragenital, G: genital, E, EG: concomitant genital, and extragenital, L: localised, S: serous, H: hemorrhagic, F: female, M: male

Case report

A 75-year-old woman presented with a generalized blistering skin rash. The 20-year-old lesions were associated with bullae, subepidermal bleeding, intermittent itching, and pain. She stated that they had developed at her hysterectomy scar 2 years postoperatively. Cutaneous examination revealed generalized poikilodermatous eruptions with hyper- and hypopigmentation, atrophy, telangiectasias, and whitish atrophic plaques with wrinkled surfaces. Moreover, keratin plugs were observed on the trunk and the upper and lower limbs, but not on the hands, feet, and mucous membranes (Figure 1A–C, 2A–2C, 3–C). Brown patches and serous and hemorrhagic bullae were scattered and superimposed on the previous lesions, causing the poikiloderma (Figure 2A, 2B, 3B). Interestingly, a serpiginous, linear, crusted ulcer had developed on the medial and posterior aspects of the left leg, 4 months before our consultation (Figure 3A, 3C).

The patient had undergone two skin biopsies in 2018 and 2019, i.e., 5 and 4 years before presentation, respectively. However, no definite diagnosis had been established until we diagnosed her with extensive EGBLS. The differential diagnoses were as follows: poikiloderma vasculare atrophicans (rejected owing to the presence of blisters and the delayed onset), poikilodermic mycosis fungoides (rejected owing to the prolonged blistering and associated scarring), post-radiation dermatitis (rejected based on the absence of a history of irradiation), graft-versus-host disease (rejected owing to the lack of a history of bone marrow transplantation), porphyria cutanea tarda or bullous lupus erythematosus (rejected because of inconsistencies with distribution of blisters and scar morphology). The subepidermal hemorrhaging detected during the biopsy in October 2018 led to confusion, because it was interpreted as an angiokeratoma and exhibited hyperkeratosis, focal parakeratosis, and atrophic epidermis. This was re-evaluated and found to be consistent with bullous LS. The findings from the second biopsy in 2019 were consistent with LS. The patient received oral methotrexate (22.5 mg) weekly, topical clobetasol propionate (0.05%) ointment twice daily, and oral prednisolone (20 mg) on alternate days. This resulted in a decrease in the lesion size and improvement in the condition.

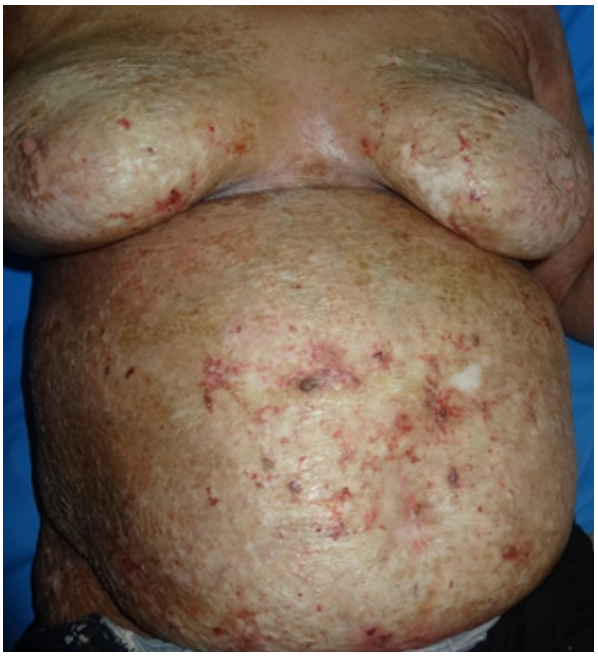

Figure 1a Porcelain-white plaques on both Breasts with atrophic wrinkled surface, hyper- and hypopigmented patches, intermingled with ecchymoses, serous and hemorrhagic bullae on the abdomen.

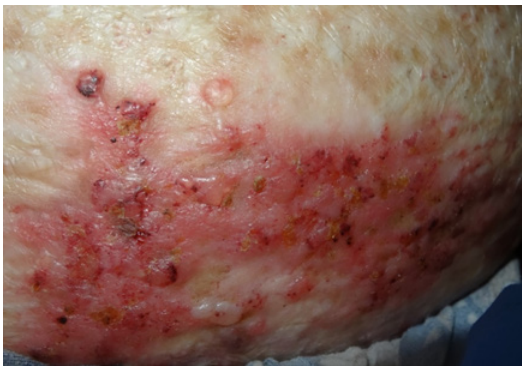

Figure 1b Close-up view of the lower abdomen: poikiloderma, serous and hemorrhagic bullae, and crusts.

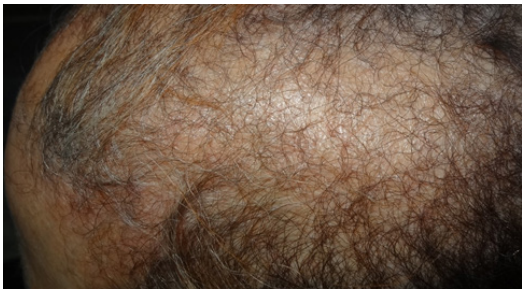

Figure 1c Diffuse scarring alopecia over the left parietal area.

Citation: Alshami MA, Alshami AM, Alshami HM. Extensive isolated extragenital hemorrhagic bullous lichen sclerosus in a 75-year-old Yemeni woman: A case report. *J Dermat Cosmetol.* 2024;8(1):4–7. DOI: 10.15406/jdc.2024.08.00256

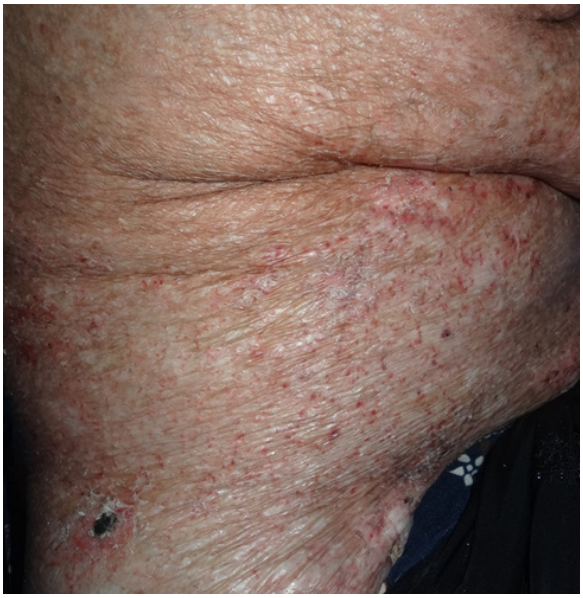

Figure 2a Lateral view of the right flank: picture of post-irradiation dermatitis, with poikiloderma, atrophic scars, telangiectasias, and atrophy.

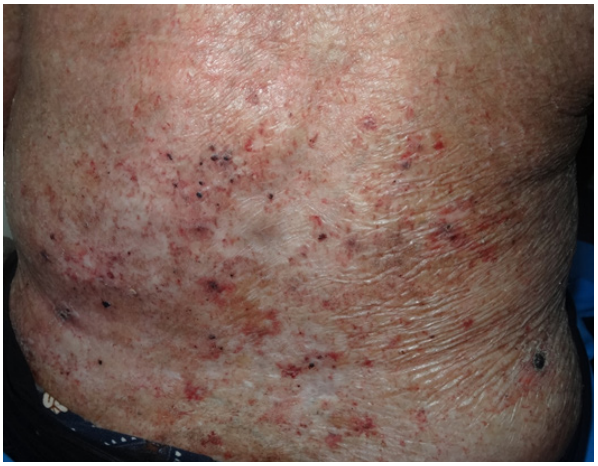

Figure 2b The lower back has wrinkled skin, atrophic white scars, ecchymoses, and hemorrhagic bullae. On close inspection, plenty of keratin plugs can be seen in the sacral area.

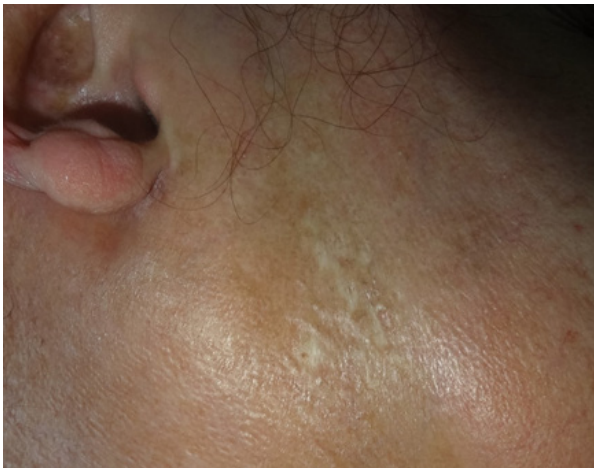

Figure 2c Whitish-yellow linear plaques over the right submandibular area.

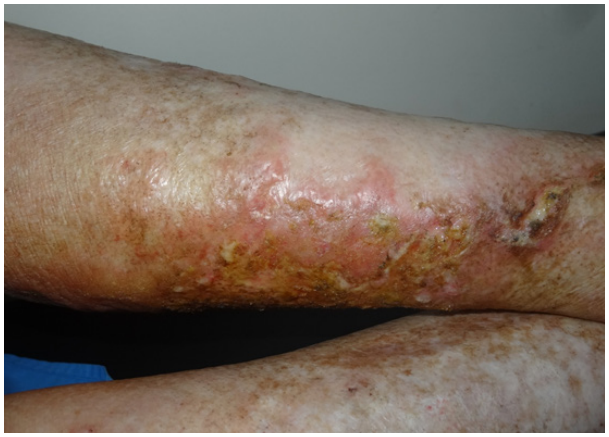

Figure 3a A linear serpiginous crusted ulcer of the left leg was seen on the anteromedial aspect.

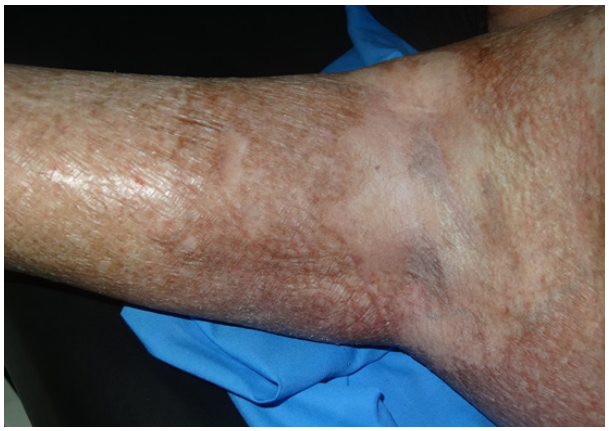

Figure 3b Right axilla, with wrinkled white skin, devoid of hairs, and adjacent poikilodermatous area.

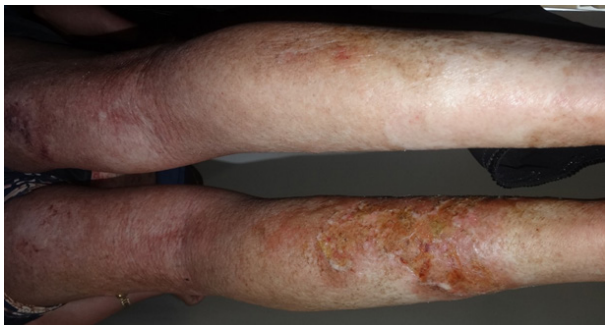

Figure 3c posterior aspect of lower limbs with involvement of both popliteal fossae and linear ulcer on the left calf.

Discussion

This case was unique for several reasons. First, the diagnosis was established 20 years after disease onset. Second, it involved >90% of the skin surface. Third, apart from the typical features, a linear, serpiginous, crusted ulcer developed on the left leg a few months before consultation. Fourth, the hands, feet, and oral and genital mucous membranes were not involved, but the face and scalp were. This case closely resembled poikiloderma vasculare atrophicans, poikilodermic mycosis fungoides, chronic graft-versus-host reaction, and post-radiation dermatitis.

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Based on clinical and histological findings, the patient was diagnosed with EGBLS. In a recent case series on the efficacy of methotrexate in cases resistant to topical treatment, prednisolone (20 mg; oral, alternate days) was administered along with methotrexate (22.5 mg; oral, weekly) and clobetasol propionate ointment (0.05%; topical, twice daily), which decreased the lesion sizes and improved the condition.[5] Unlike genital LS, EGLS is not associated with a risk of carcinomatous transformation, although some cases have been reported.[6]

Previously, LS has been reported at scar sites; in the present case, the lesions developed at the site of a hysterectomy scar on the abdominal wall 2 years after the surgery.[7] The breasts are commonly and severely affected.[4] In this case, both breasts were severely affected, with atrophic wrinkled skin and a few hemorrhagic bullae (Figure 1A).

Consistent with previous reports, the lower back and waistline were significantly affected in this case (Figure 1A, 2A, 2B).[1] Additionally, several reports have described the involvement of the abdomen, which was also observed in this case (Figure 1A, 1B).[8]

Although rarely reported, the face was bilaterally affected on the submandibular margins in this case (Figure 2C).[9]

Furthermore, only a few cases of scalp involvement in the form of diffuse scarring alopecia have been reported; this was observed in the present case (Figure 1C).[10]

Involvement of the hands and feet, which is rarely reported, was also not observed in our case. Flexures are common sites of LS; in our case, the axillae and cubital and popliteal fossae were affected and devoid of hair (Figure 3B,3C).[8] Follicular plugging is characteristic of EGLS and was observed at many sites in this case (Figure 2B).[4] This report details a rare case of EGBLS with unusual features that delayed the diagnosis for 20 years; it provides data for faster diagnosis and adequate treatment in similar cases in the future.

Acknowledgments

None.

Conflicts of interest

The authors declare there is no conflicts interest.

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