Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/18355
DOI URL: http://dx.doi.org/10.21474/IJAR01/18355

RESEARCH ARTICLE

EXTRAGENITAL LICHEN SCLEROSUS ET ATROPHICUS : A CASE REPORT

Meryem Elomari Alaoui[1], Amani Fliti[1], Ismail Boujida[2], Mariame Meziane[1], Kaoutar Znati[2], Laila Benzekri[1], Nadia Ismaili[1] and Karima Senouci[1]

1. Department of Dermatology and Venerology, University Hospital Center IbnSina, University of Mohamed V, Rabat, Morocco.
2. Department of Histopathology, University Hospital Center IbnSina, University of Mohamed V, Rabat, Morocco.

Manuscript Info

Manuscript History
Received: 28 December 2023
Final Accepted: 30 January 2024
Published: February 2024

Key words:-
Lichen Sclerosus, Atrophicus, Cutaneous, Inflammatory

Abstract

Lichen sclerosus et atrophicus (LSA) most commonly presents as atrophic plaques in the genital region but rarely can occur in extra-genital locations (1), reportedly present in 6% to 20% of patients. It's a pathology that can be underdiagnosed and under-treated. The diagnosis should be confirmed with a skin biopsy, and early, thorough treatment should be initiated.(2)We report a case of lichen sclerosus et atrophicus in a woman with purely cutaneous involvement.

Introduction:

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory skin disease commonly affecting the anogenital areaand predominantly female. However, the cutaneous involvement remains less known and studied. (3)

We report a case of lichen sclerosus et atrophicus in a woman with purely cutaneous involvement, highlighting the importance of making this diagnosis in the absence of obvious genital involvement.

Case Report:
A 41-year-old woman with a history of hypothyroidism and diabete under treatment who had for 8 years, dépigmented lesions slightly pruriginous, gradually increasing in size, initially affecting the back and trunk, then extending to the lower limbs.

Clinical examination showed multiple macules, slightly sclerotic in the centre, atrophic in places, 1 to 5 mm in size, ivory-white in colour and confluent in patches on the back, trunk and lower limbs. (figure 1-2)

Oral and genital mucosa were normal. The rest of the clinical examination was normal.

However, histology showed an epidermal atrophy associated to basal vacuolization and moderate dermal fibrosis with lymphocytic infiltrate orienting to the diagnostic of cutaneous LSA. (Figure 3)

Complete blood count, routine blood biochemistry, antinuclear antibody (ANA) and hepatitis B and C serologies were within normal limits.

Corresponding Author:- Meryem Elomari Alaoui
Address:- Department of Dermatology and Venerology, University Hospital Center Ibn Sina, University of Mohamed V, Rabat, Morocco.

The patient was initially treated by dermocorticoids for3 months, and she did not shows any improvement, and then later she was treated with methotrexate at a dose of 25mg/week with an emollientand there was slight improvement of the lesions on back. A close follow up was planned.

Discussion:

LSA was first described by Hallopeau in 1887 and it received multiple names such as kraurosis vulvae, balanitis xerotica obliterans, white spot disease, leukoplakia and lichen sclerosus et atrophicus. The final term “lichen sclerosus” was accepted in 1976 by the International Society of the Study of Vulvovaginal Disease (5-6)

Lichen sclerosus (LSA) is an underdiagnosed inflammatory mucocutaneous condition affecting the anogenital areas. Purely extra-genital localization is only seen in 2.5% of cases (7). Extragenital atrophic lichen sclerosis occurs mainly in post-menopausal women (8), mainly in the submammary region, neck, shoulders, inner thighs, wrists and upper back (9). The pathophysiology may involve several factors: hormonal, genetic (HLA II DQ7), infectious (Borrelia burgdorferi) or traumatic (Köebner phenomenon).(10)

Clinically, extragenital LSA is asymptomatic, the lesions take the form of whitish or pearly-white, "porcelain-like", atrophic patches, mainly affecting the trunk, back, roots of limbs and folds. Pruritus is inconsistent.

Blaschkolinear and bullous clinical forms have been described [11,12]. Diagnosis is based on skin histology, which reveals atrophy of the squamous epithelium with horizontalization of the basal layer, follicular hyperkeratosis, and above all the presence of a subepithelial band of fibrous or oedematous collagen in the superficial dermis, devoid of elastic fibres when stained with orcein.(13)

The treatment of CLS is not codified and consists of local treatment (potent topical DC or intralesional corticosteroid, calcipotriol, tacrolimus), more or less combined with general treatment (synthetic antimalarials, retinoids, general corticosteroid therapy, methotrexate or ciclosporin) depending on the course of the disease under local treatment, and UVB phototherapy can be also used. (14)

Conclusion:

Extragenital lichen sclerosus et atrophicus is a rare disorder that can be difficulte to differentiate clinically from a number of disorders of hypopigmentation and sclero-atrophic disorders, and hence requires skin biopsy.Unlike genital LS, extragenital LS is not associated with a risk of carcinomatous transformation.

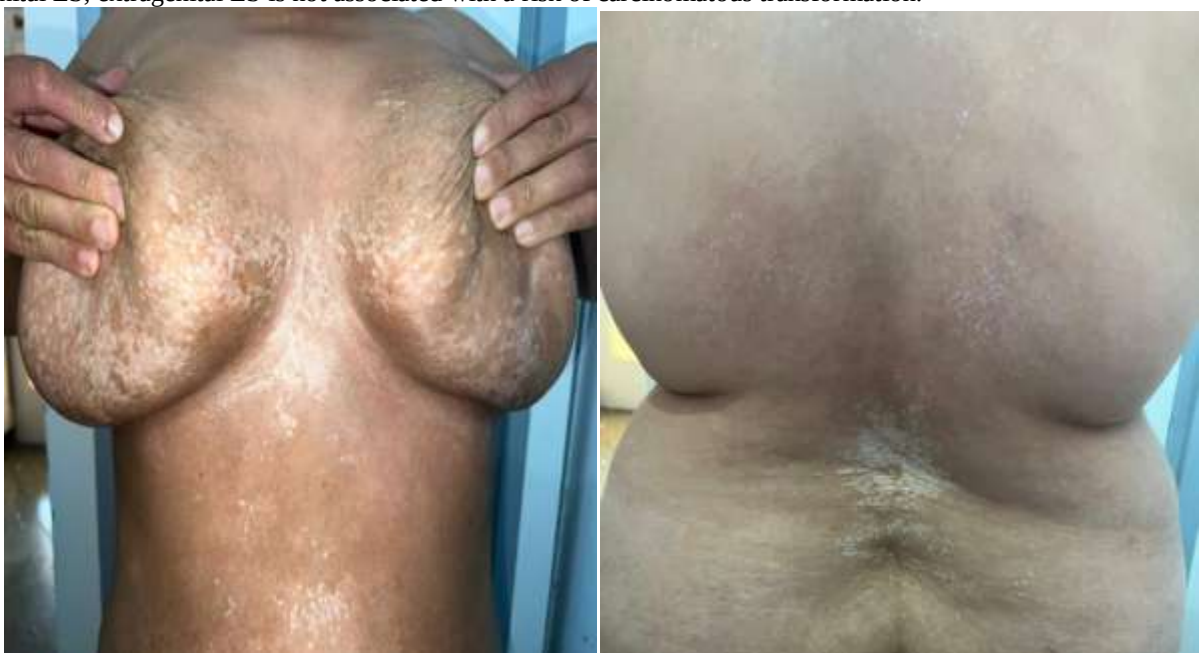

Figure 1-2:-Clinical image shows multiple ivory-white macules, slightly sclerotic in the center, atrophic in spots on the back and trunk.

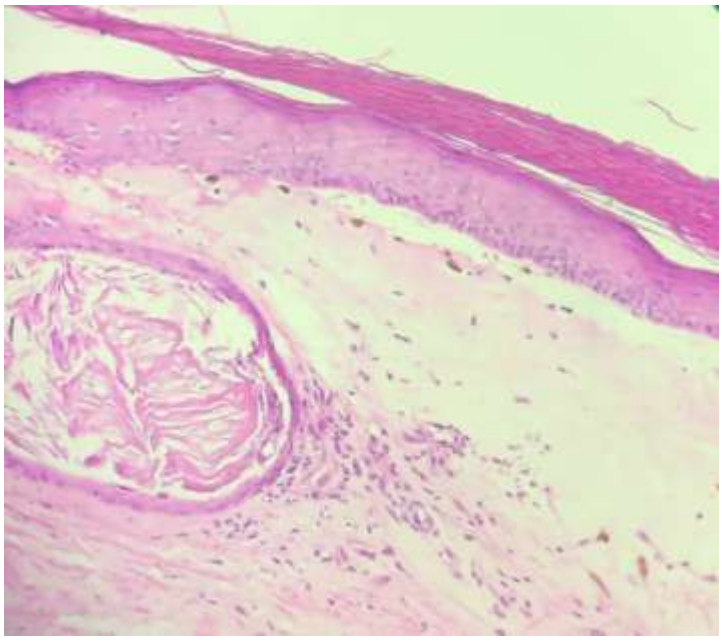

Figure 3: Histological image of lichen sclerosus et atrophicus.

Reference:-

1. Yasar S, et al. A case of lichen sclerosus et atrophicus accompanying bullous morphea. Ann Dermatol 2011; 23:S354
2. Kirtschig G. Lichen Sclerosus-Presentation, Diagnosis and Management. DtschArztebl Int. 2016 May 13;113(19):337-43. doi: 10.3238/arztebl.2016.0337. PMID: 27232363; PMCID: PMC4904529.
3. Burshtein, A., Burshtein, J. &Rekhtman, S. Extragenital lichen sclerosus: a comprehensive review of clinical features and treatment. Arch Dermatol Res **315**, 339–346 (2023). https://doi.org/10.1007/s00403-022-02397-1
4. De Luca DA, Papara C, Vorobyev A, Staiger H, Bieber K, Thaçi D, Ludwig RJ. Lichen sclerosus: The 2023 update. Front Med (Lausanne). 2023 Feb 16;10:1106318. doi: 10.3389/fmed.2023.1106318. PMID: 36873861; PMCID: PMC9978401.
5. Hallopeau H. Leçons cliniques sur les maladies cutanées et syphilitiques. Union Med Can. (1887) 43:472. [Google Scholar]
6. Kaufman RH, DiPaola GR, Friedrich EG, Jr, Hewitt J, Woodruff JD. New nomenclature for vulvar disease. J CutanPathol. (1976) 3:159–61. doi: 10.1111/j.1600-0560.1976.tb01105.x, PMID: [PubMed] [CrossRef] [Google Scholar]
7. Benhiba H, et al. Lichen scléro -atrophiqueextra-génital. Presse Med. 2015: 1-3.
8. 27. Arif T, Fatima R, Sami M. Extragenital lichen sclerosus: a comprehensive review. Australas J Dermatol. (2022) 63:452–62. doi: 10.1111/ajd.13890. [Epub ahead of print], PMID: [PubMed] [CrossRef] [Google Scholar]
9. 98. Powell JJ, Wojnarowska F. Lichen sclerosus. Lancet. (1999) 353:1777–83. doi: 10.1016/s0140-6736(98)08228-2 [PubMed] [CrossRef] [Google Scholar]
10. Ballester I , Bañuls J , Pérez-Crespo M, Lucas A. Extragenital bullous lichen sclerosusatrophicus. Dermatol Online J. 2009; 15(1): 6. PubMed | Google Scholar
11. Choi SW, Yang JE, Park HJ, Kim CW. A case of extragenital lichen sclerosus following Blaschko's lines. J Am Acad Dermatol. 2000;43(5 Pt 2):903–4. [PubMed] [Google Scholar]
12. Khatu S, Vasani R. Isolated localised extra-genital bullous lichen sclerosus et atrophicus: a rare entity. Indian J Dermatol. 2013;58(5):409. [Article PMC gratuit] [PubMed] [Google Scholar]
13. Cavelier-Balloy B. Lichen scléreux. Ann Dermatol Venereol. 2012;139(1):65–7. [PubMed] [Google Scholar]
14. Cuellar-Barboza A, Bashyam AM, Ghamrawi RI, Aickara D, Feldman SR, Pichardo RO. Methotrexate for the treatment of recalcitrant genital and extragenital lichen sclerosus: A retrospective series? Dermatol Ther. 2020;33:e13473. doi: 10.1111/dth. 13473. [PubMed] [Google Scholar].