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Linchen Sclerosus Et Atrophicus: A Case Study

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ABSTRACT

Background: Lichen sclerosus is a chronic inflammatory skin disorder that most commonly affects women before puberty or after menopause. Lichen sclerosus is characterized by skin changes of the external genitalia. The most common distribution is involving the vulva and perianal area. The head of the penis and other parts of the body may also be affected. In fact, this skin condition can affect any skin surface with the most common extra-genital sites being on the trunk, a limbs.[1] Extragenital involvement is found in approximately 11% of women and is rare in men. Extragenital involvement alone is uncommon and has been reported in about 6% of all women with LSA. The aetiology of LSA is unclear, but genetic, physiological and environmental factors are thought to be involved. **Objective:** To describe the clinical features and complications of disease and discuss the current management. **Methods:** Detailed history, physical examination and histopathology findings, a diagnosis of generalized lichen sclerosus Et atrophicus (LSA) was made. Other laboratory examinations were unremarkable. **Conclusion:** Generalized type is rare and genital involvement is most frequent and often the only site of involvement. As there is no standard treatment for LSA, the patient received various treatments for topical steroid, tacrolimus and narrow –band ultraviolet B therapy. The skin lesions has softened and its color improved after treatment. As, this case is reported an uncommon type of LSA with generalized hyper pigmented and sclerotic skin lesion in a postmenopausal female patient.

Keywords: Generalized, Lichen sclerosus Et atrophicus.

INTRODUCTION:

Lichen sclerosus is a chronic inflammatory skin disorder that most commonly affects women before puberty or after menopause. Although rare, it can also be seen in men. When found in males, the disease is known as balanitis xerotica obliterans.

Lichen sclerosus is characterized by skin changes of the external genitalia. The most common distribution is involving the vulva and perianal area. The head of the penis and other parts of the body may also be affected. In fact, this skin condition can affect any skin surface with the most common extra-genital sites being on the trunk, a limbs.[1] Extragenital involvement is found in approximately 11% of women and is rare in men. Extragenital involvement alone is uncommon and has been reported in about 6% of all women with LSA. The aetiology of LSA is unclear, but genetic, physiological and environmental factors are thought to be involved.[2]

Moreover, an autoimmune basis has been proposed for the etiopathogenesis of LSA. Two separate population-based studies of genital LSA noted an increased prevalence of psoriasis in patients with LSA, 17.0% and 7.5%, respectively, compared with a 1.5%–2.5% incidence in the general population. Recently, autoantibodies against extracellular matrix protein 1 have been proposed to play a role in the pathogenesis of LSA. Along with this, a genetic basis has been proposed with positive family history in 12% of cases and is seen in identical and nonidentical twins. An association with human leukocyte antigen Class II antigen DQ7 and interleukin-1 receptor antagonist gene polymorphisms has been identified in several studies. Furthermore, infection with spirochetal microorganisms such as *Borrelia burgdorferi* and atypical mycobacterial infections have been reported. Human papillomavirus (HPV) and hepatitis C virus have also been implicated as possible causative agents. LSA also shows Koebner's phenomenon at sites of trauma, burns, radiotherapy, and vaccination. It can also occur post vulvectomy and in episiotomy scars. Inspite of this, hormonal factors, atopy, allergic contact dermatitis, obesity and anatomical abnormality can be the contributing factor for LSA.[3]

Clinically, LSA is characterized by papules and plaques of porcelain-white atrophic or thickened skin that may become purpuric, blistered or eroded. The genital lesions are pruritic whereas the extragenital ones, which usually arise on the inner thigh, submammary area, neck, shoulders and wrists, are asymptomatic.[2] The extra-genital cutaneous lesions of LSD consist of flat - topped, white macules that tend to coalesce to form white patches. Often, small, comedo-like follicular plugs are seen on their surface.[3] In the later stages, atrophy occurs and the surface of the lesions becomes wrinkled and may actually be depressed. Some patients with lichen sclerosus do not have any symptoms, whereas others experience intense itching, discomfort and/or

erosions/ulcers. Lichen sclerosus typically has a remitting relapsing course that is complicated by permanent scarring of the affected areas. The disorder is not contagious nor is it a sexually transmitted disease.[4]

Histologically, LSA has a characteristic pattern. Hyperkeratosis, follicular occlusion, thinning of the epidermis, and vascular alterations in the basal layer are seen in the epidermis. There is a large area of subepidermal edema with homogenization of collagen, sclerosis, and dilation of the small vessels with hemorrhage. A diffuse perivascular infiltrate of lymphocytes appears under the edema in the middle third of the dermis. Reticular dermal changes of fibrosis and inflammation in morphea is in contrast with edema and loss of elastic tissue in LS, which helps to differentiate both conditions.

The rate of spontaneous resolution may be lower than 25%. Currently, there is no effective treatment available for LS. Most patients are initially treated with potent topical corticosteroids. Various treatments including PUVA, topical testosterone and estrogen, topical tacrolimus or pimecrolimus, antimalarial agents, penicillin, topical retinoids, and vitamins, have been tried. NBUVB has both anti-inflammatory and immunosuppressive effects. Several studies have demonstrated that both UVA1 and NBUVB increase matrix-metalloproteinase levels in human skin and cultured dermal fibroblasts, which explains the effectiveness of UVA in sclerosing skin diseases, but LSA affects only the epidermis and superficial dermis, so along with UVA, NBUVB is also effective in LSA.[5]

Case report

A 63 years old female visited Outpatient department at Guru Gobind Medical College and Hospital, Faridkot with history of Mastectomy in 1998 and chief complaints of multiple whitish lesion under breast and upper back found a year ago. Physical examination revealed that there is also dark coloured lesion in pelvic area. There lesions are characterized multiple hypo pigmented atrophic macules present inframmary area and upper back along with inability to pinch overlying skin in pelvic area. Firstly, dermatologist diagnosed the patient with provisional diagnosis i.e. Morphea and suggested to go for Skin biopsy at pelvic area. The skin biopsy test reports revealed from H & E stained sections examined for skin biopsy is comprised of epidermis and dermis. Epidermis shows the hyperkeratosis, hydropic degeneration of basal layer with atrophy of stratum malpighi and mild spongiosis along with that the dermis layer also shows the homogenization of cell bundle which proved the Lichen Sclerosis et atrophicus (extragenital). Other laboratory examinations were unremarkable. She began treatments that consisted of systemic and topical steroids. Because there is no definite treatment guideline for generalized LSA, various treatments had been attempted, including topical steroids, tacrolimus. Along with that she also referred another dermatologist and he had prescribed methotrexate to the patient which was stopped because of hematesis but she is now on topical steroids but this disease further spreads to other area. We received the patient consent form about publishing all photographic material.

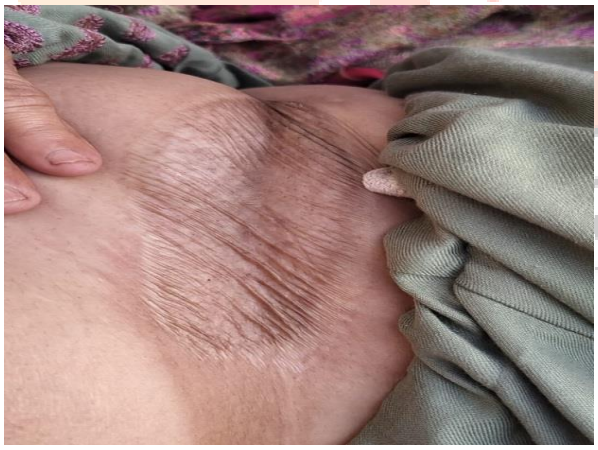

Figure 1: Depigmentation and scaly erythema at pelvic area

DISCUSSION

In both male and female, LSA mainly occurs in the genital area and sometimes accompanies symptoms such as itching and dysuria. When extragenital areas are involved, most cases show localized invasion, and generalized involvement affecting more than 2 anatomic regions is very rare. Our patient manifested with characterized multiple hypo pigmented atrophic macules present inframmary area and upper back along with inability to pinch overlying skin in pelvic area. In some previously reported cases, systemic blisters or ulcerations were accompanied by generalized LSA. Our case is thought to be a rare case of generalized LSA, as blisters or ulcerations were not observed. In addition, preceding studies about LSA hypothesized that this may be related to autoimmune factors because 21.5% of patients had autoimmune diseases and 42% had autoantibodies. However, there was no specific laboratory finding in our case.

Histopathologically, Epidermis shows the hyperkeratosis, hydropic degeneration of basal layer with atrophy of stratum malpighi and mild spongiosis along with that the dermis layer also shows the homogenization of cell bundle. Morphea should be differentiated from LSA. Morphea shows relatively normal epidermis without follicular plugging and does not include subepidermal separation; fibrosis is present with collagen and elastic fibers at the dermis. Inflammation of the sub cutis can also be observed. According to previous reports on generalized LSA, many cases were accompanied by morphea. In the present case, the histopathological findings were consistent with LSA and did not show any features of morphea.

In addition, this case is meaningful because the patient is not improved through treatments including topical steroids and tacrolimus. But, there have been reports about successful treatments of genital LSA with topical steroids and tacrolimus, there is no established treatment guideline for generalized LSA. As our case showed no alleviation of symptoms and improved skin lesions, but the patient had not gone for narrow band UVB along with tacrolimus and topical steroids that can be the reason this treatment might not be effective. However, future studies are needed to confirm this result.[6]

REFERENCES:

1. Lichen Sclerosus- Symptoms, Causes, Treatment .National Organisation for Rare disorders. http://rarediseases.org> Rare Diseases.

2. Guerriero C et al. extragenital lichen sclerosus and atrophicus treated with topical steroids and retinoids in a child with vitiligo. International Journal of Immunopathology and Pharmacology.2008;21(3):757-759.

3. Marfatia Yogesh, Surani Asthma and Baxi Reema. Genital Lichen sclerosus et atrophicus in females. An update.Indian J Sex Transm Dis AIDS.2019;40(1)6-12.

4. Lagace Francois and Jacobe Heidi. Lichen Sclerosis. 2023. [cited 2024 April 4]. Avalibale from. http://rare diseases.org> Rare disease.

5. Diwan G. Nilofar and Nair A Pragya. Extragenital Lichen Sclerosus et atrophicus along the lines of Blaschko. Indian Dermatol Online J. 2015 Sept-Oct; 6(5): 342-344. Avalibale from. http://www.ncbi.nlm.nih.gov>articles>PMC4594396.

6. Hong et al. A Case of Generalised Lichen Sclerosus et Atropicus. Ann Dermatol. 2020 Aug; 32 (4) : 327-330.